



CRANIOFACIAL SURGERY

EDITED BY

SETH R. THALLER
JAMES P. BRADLEY
JOE I. GARRI

WITH AN INTRODUCTION BY
HENRY K. KAWAMOTO



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New York London

Informa Healthcare USA, Inc.
52 Vanderbilt Avenue
New York, NY 10017

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Printed in the United States of America on acid-free paper
10 9 8 7 6 5 4 3 2 1

International Standard Book Number-10: 0-8493-8210-6 (Hardcover)
International Standard Book Number-13: 978-0-8493-8210-9 (Hardcover)

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Library of Congress Cataloging-in-Publication Data

Craniofacial surgery / edited by Seth R. Thaller, James P. Bradley, Joe I. Garri.
p. ; cm.

Includes bibliographical references and index.

ISBN-13: 978-0-8493-8210-9 (hb : alk. paper)

ISBN-10: 0-8493-8210-6 (hb : alk. paper)

1. Skull--Abnormalities--Surgery. 2. Face--Abnormalities--Surgery. I. Thaller, Seth R.

II. Bradley, James P., 1965- III. Garri, Joe I.

[DNLM: 1. Craniofacial Abnormalities--surgery. 2. Facial Bones--surgery. 3. Skull--surgery.

WE 705 C89068 2007]

RD763.C7512 2007

617.5'2059--dc22

2007026349

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This book is dedicated to my mentors, who have served as advisors, counselors, guides, teachers, and gurus; to my family, who has encouraged my ambitions and provided loving support; and to my greatest mentor, my father, who recently passed away.

—SRT

This book is dedicated to Henry K. Kawamoto, MD, DDS. He taught his craniofacial fellows and other students to strive for excellence in surgical care for craniofacial patients by following accepted principles and testing new techniques. Dr. Kawamoto fostered the importance of mentoring surgeons to ensure optimal care for future patients.

The following invited speakers for the Kawamoto Lectureship are a tribute to this ongoing propagation of knowledge: Paul L. Tessier, MD (1998), Hugo L. Obwegeser, MD, DMD (2000), Fernando Ortiz-Monasterio, MD (2002), Daniel Marchac, MD (2004), S. Anthony Wolfe, MD (2006), and Joseph G. McCarthy, MD (2008).

—JPB

For those of us who have had the pleasure and privilege to study under Dr. Kawamoto, the craniofacial fellowship year has been the crown jewel of our long educational careers. This book is our way of expressing our gratitude to such a pivotal figure in our professional lives. It is also our way of continuing his legacy and bringing his teachings to current and future craniofacial surgeons who did not experience them first hand.

—JG

Introduction

The UCLA Craniofacial Fellows represent a special breed.

Years ago, Mary McGrath, then Chief of Plastic Surgery at George Washington University, asked, "How do you keep turning out outstanding fellows?" The simple answer was, "They were outstanding before they started." My task was to stay out of their way. Their job was to keep me out of trouble as I showed them a few things.

Amongst themselves, the Fellows share a special and unique esprit de corps. As their mentor, I can attest to this. They represent the joys of teaching.

The word mentor is derived from Greek mythology and the epic tale, *The Odyssey*. However, the Fellows' accomplishments are no myth. Each has developed a successful practice in plastic surgery. And, as is the hope of all mentors, they have superceded their training.

I am anxious to read their thoughts. Hopefully, you, as readers, will find the book equally rewarding.

Henry K. Kawamoto



Foreword

I first met Henry Kawamoto when we were general surgery residents at the Columbia-Presbyterian Medical Center. I recall Henry already knew he wanted to pursue a career in plastic surgery of the face. It is, however, unlikely that he had heard of the term “craniofacial surgery” because we actually met in 1967, the same year of Tessier’s landmark paper at the International Congress in Rome.

Following our year as chief resident in general surgery, we continued together, albeit only a few miles south in Manhattan, as residents under the late John Marquis Converse at the Institute of Reconstructive Plastic Surgery at the New York University Medical Center. This was an outstanding educational experience. Converse was dynamic and enthusiastic. Incredibly academic and creative, he was especially interested in orbital surgery and the correction of orbital hypertelorism through the “craniofacial” route. During Henry’s first year as a plastic surgery resident, Paul Tessier was the visiting Kazanjian Professor at New York University and he made an enormous impact on a standing-room-only audience, including our young plastic surgery resident from California.

I would suspect that it was Henry’s first meeting with Tessier and the beginning of an especially close professional collaboration that has extended over 35 years. Following graduation from the Institute, Henry spent a year with Tessier at L’Hôpital Foch and was one of the first surgeons to bring the Tessier concepts and techniques to the United States. He initiated and developed an internationally acclaimed craniofacial surgery service at University of California Los Angeles. Not only has he developed innovative techniques for the correction of congenital and post-traumatic craniofacial deformities, but also began an outstanding year-long fellowship in craniofacial surgery that has produced many of the leaders of the “third generation” of craniofacial surgeons, Henry being of the second and his teachers, Converse and Tessier, being of the first. And so, it is only fitting that the surgical lineage, or the “craniofacial surgical DNA,” is continued in the chapters of this text by the graduates of his fellowship program under the talented editorship of James Bradley.

How proud Converse and Tessier would be of this literary achievement! It demonstrates that their pedagogical efforts have been rewarded and that their professional progeny have built on their contributions and advanced this surgical discipline for the betterment of all patients with craniofacial deformities.

As a friend and colleague over these years, I salute Henry for all he has accomplished, not only for his skill and innovation in the operating room but also for his contribution to our meetings. What a tribute to him the graduates of his fellowship program have provided in compiling this outstanding text on state-of-the-art craniofacial surgery.

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Preface

Craniofacial surgery is a distinct subspecialty of plastic and reconstructive surgery that has witnessed tremendous advancements over the last decade. The origins of craniofacial surgery derive from plastic surgeons' experience correcting extensive traumatic injuries during the First World War. However, the specialty was most influenced by the pioneering vision of Dr. Paul Tessier of Paris, France, who described a combined intra- and extracranial approach in the 1970s, and who had the foresight to employ these techniques for the correction of extensive congenital and acquired deformities. From this early work, and the eventual diaspora of his pupils such as Dr. Henry Kawamoto, craniofacial surgery is now recognized as a distinct and desired sub-specialty within the veil of the broad field of plastic surgery. The role has continued to expand, and craniofacial techniques are now frequently used in elective cosmetic surgery procedures as well as deformity correction.

The goal of this book is to exemplify and demonstrate contemporary techniques and general concepts employed in the sub-specialty of craniofacial surgery. The main tenets of the field of craniofacial surgery center around skeletonization, three dimensional mobilization, anatomic re-positioning and rigid stabilization of the bony skeleton, and autogenous augmentation and/or replacement. This book will provide a forum for past University of California Los Angeles fellows who have had the once-in-a-lifetime opportunity to learn their basic skills and study under the tutelage of the renowned Dr. Henry Kawamoto, a.k.a. "the Samurai." As they have become leaders in the field of plastic surgery, each of these fellows has had the opportunity to expand upon this foundation and to further develop his or her own clinical and research interests within this ever-changing discipline. We are pleased to share this with our colleagues in plastic and reconstructive surgery and to describe the standard of care for managing these challenging clinical entities.

Acknowledgment

We would like to acknowledge the assistance from Rebekah Ashley, who provided us with the energy and direction to complete this task.

*Seth R. Thaller
James P. Bradley
Joe I. Garri*

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A History of Craniofacial Surgery Seen Through a UCLA Prism

S. Anthony Wolfe

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Paul Louis Tessier, a native of Nantes, France, was working on the burn unit at Hôpital Foch in Boulogne, on the other side of the Bois de Boulogne from Paris, in the late 1950s and 1960s. Hôpital Foch had been built after the First World War, largely with American donations [in particular from the Singer (sewing machine) family], and continues today as a major teaching hospital in France. Originally, Tessier had been part of the Maxillofacial Service headed by Maurice Virenque, who had been his Chief when he was working in a Military Hospital in Le Mans. There were, for a time, two maxillofacial units at Foch: one headed by Virenque, and the other by Gustave Ginestet, a politically connected and somewhat self-inflated individual who held the rank of General in the French Army. Virenque, Tessier's patron, died, leaving Ginestet in control of the only maxillofacial service at Foch. Ginestet was not fond of Tessier (perhaps he was threatened by him?), and he issued an edict to the dental laboratory that they not under any circumstances provide support to Tessier.

Possibly this animosity dated back to the time of the seaplane crash of Jacqueline Auriol, the daughter-in-law of the President of France, in 1949. She sustained a Le Fort III fracture, and, according to a conversation I had with Tessier one evening over several very good bottles of Bordeaux (Petrus, I recall, 1982), none of the French surgeons called in to consult on the illustrious aviatrix had much of an idea of what to do. A well-known Parisian rhinoplastic surgeon had been consulted, and he suggested...a rhinoplasty (even though the patient still had a floating mid-face!). The family was told by someone that they should ask Tessier to see the patient, and this was eventually arranged, even though the patient was in the service of Dr. Ginestet.

After Tessier had seen the patient, he told the family that, in his opinion, a bone graft would be necessary to provide consolidation of the non-united fracture. Ginestet was able to prevent Tessier himself from performing the operation and getting any political capital out of it by saying that the only person who could perform such a bone graft was John Marquis Converse of New York. Converse had many connections in France because his father had been the Medical Director of l'Hôpital Americaine in Paris, and he had gone to medical school in France and received some of his surgical training there. Converse did indeed end up caring for the famous daughter-in-law, and the Auriol family helped in the formation of the Institute for Reconstructive Plastic Surgery at New York University (NYU) with a generous donation.

So, Tessier's only position in the late 1950s at Foch was on the burn unit. At some point in the 1950s, Tessier began seeing patients with Crouzon disease. He knew about the patient that had been operated upon by Sir Harold Gillies, first in 1942, and then again, after considerable relapse, in 1949 (Hugo Obwegeser was the assistant on the second operation, where ox cartilage was implanted to camouflage the persistent undercorrection).

Tessier realized that bone grafts would need to be placed in the surgically created gaps to prevent relapse. He also designed osteotomies that differed from those of Gillies (for instance, instead of cutting across the back of the hard palate, Tessier performed a pterygo-maxillary disjunction). These osteotomies were tested on cadaver heads in the anatomy department of Tessier's old medical school in Nantes, where he would go on the train on evenings and weekends after finishing his very heavy surgical schedule in Paris. Finally, he performed his

first Le Fort type III in 1958, at Hôpital Foch (Tessier insists it should not be called a Le Fort III osteotomy, since the pterygoid plates are kept intact). The patient was an adult with Crouzon disease named M. Anquetil, with a quite severe deformity. Tessier performed the osteotomies through limited facial and oral incisions (the coronal incision was not in use at that time), advanced the midface over 25 mm, and inserted iliac bone grafts into the surgically created gaps. The result, Tessier felt, was not sufficiently stable, so he had a rapid consultation with his instrument makers and had them make a “diadem,” or head frame, which would maintain the midface in the position he wished post-operatively. (I have this diadem in a drawer in the office, and should send it on with Tessier’s notes on Monsieur Anquetil to the Plastic Surgery Archives in Boston.)

This operation was repeated many times over the subsequent decade, now through a coronal approach with a number of clever self-retaining osteotomies (recall that Tessier was still denied access to the dental laboratory). And, in 1967, at the International Meeting in Rome, after Tessier was convinced that he had developed a safe and reliable procedure, the Le Fort III osteotomy was presented.

All present at that meeting were shocked, to say the least, and many arranged to either come to Paris to see Tessier operate, or send a junior colleague from their unit.

I was able to hear Tessier’s first presentation in North America, at the ASPRS (ah, the good old days, when we still had that “R”) meeting in Montreal. Like those who had been in Rome, I went away awed, and decided that I wanted to go to Paris too. I was at that time in my last year of general surgical residency at the Peter Bent Brigham Hospital in Boston, and had already decided to go into plastic surgery. One of the perks of the Brigham residency at that time was a travel fund that allowed the senior resident to take several weeks of travel anywhere he wished in the United States.

In 1972, as a plastic surgery resident, I went to Chicago, where Sam Pruzansky had invited Tessier to come operate. This was the first time I met Tessier in person and, probably because of the fact that I was with Millard, who Tessier knew and had visited, he accepted that I come to Paris after I finished in Miami.

At about the same time, Henry K. Kawamoto, Jr., had finished his general surgical training at Columbia (a co-resident there was Joe McCarthy, who had been good friends in medical school with John Mulliken, who went on to the Mass General for his general surgery training, then to Baltimore for plastic surgery, and finally to Boston Children’s, where he collaborated over many years with Paul Tessier).

Henry “the K” decided on the NYU program. (I forget what the middle initial “K” stands for, but anyone who has skied with Henry might think it would be Kawasaki or Kamikaze.)

Tessier was invited to be the Kazanjian Professor at NYU in 1971, and he performed several operations there with Converse. Francoise Firmin, who had training with Tessier in Paris, was spending a year at NYU at that time. Henry Kawamoto was not there then, however, since apparently he was taking a vacation in Bermuda at the time of the Tessier visit. Converse later did arrange for Henry to go to Paris, perhaps somewhat as a spy to find out what new procedures Tessier was up to, and Henry and Kathy packed up and went to Paris in 1974.

I arrived in Paris in July of 1974, having missed Henry by a month or so. Elizabeth Hecht, Tessier’s legendary scrub nurse, had been quite fond of Henry, and I finally got to meet him in Chicago in 1975, where one of the first meetings on craniofacial surgery was arranged by Sam Pruzansky.

Henry had gone back to Los Angeles, and the new full-time Chief who took over after Franklin Ashley was Harvey Zarem, an Edgerton trainee. Harvey was apparently unwilling to have Henry join the UCLA faculty unless he took a full-time position, which Henry was reluctant to do. Only after the intercession of Tessier did Harve allow Henry to have a part-time faculty appointment.

After my return from France, Henry and I became good friends. For a decade or so we gave a Teaching Course at the ASPRS meeting on orthognathic surgery, which was a sell-out for a number of years, but eventually attracted only about a dozen or so registrants (either interest in the subject had flagged, or everyone who had an interest in the subject had already taken the course).

Between 1975 and 1995, Tessier operated at various centers in the United States for between one to two months a year, and did only craniofacial work when he was here. (In Paris, although the operative schedule was gargantuan—10 to 15 cases a day, six days a week—about half of the schedule was garden variety plastic surgery, including cosmetic.) Besides New York, Philadelphia, and Dallas on only a few occasions, there was also Los Angeles (with Henry), San Francisco, and Louisville, and repeated trips to Kansas City, Houston, Norfolk, and Boston. (Tessier over the years probably did more craniofacial surgery in the United States than any American surgeon.) Spending a few days with Dr. Tessier during these visits was truly continuing medical education in craniofacial surgery, for here we could see the specialty growing before our eyes, since virtually every procedure we use in craniofacial surgery was developed by Dr. Tessier. The use of the cranium as a bone graft donor site, the monobloc frontofacial advancement, and the facial bipartition were put into common usage by Tessier after our time with him in Paris.

On a number of occasion, Henry and I went with Tessier to UNICAMP (University of Campinas) where our good friend Cassio Raposo do Amaral was developing a first rate craniofacial unit (SOBRAPAR). There was a limitless reservoir of pathology there, and a host of techniques were developed in what I thought of as the Aberdeen Providing Ground of craniofacial surgery. I well recall a young girl named Sossia, who had major craniofacial deformities following an automobile accident and several traditional, but ineffective, attempts to correct them. Tessier removed a large block of methylmethacrylate from her forehead, and there was a sudden gush of clear fluid. He stopped for a moment, peering downward, and then inserted a rugine (periosteal elevator) to its hilt through the defect in the forehead. He then turned to us and said he was pleased to report that he was able to see the foramen of Munro. A temporalis muscle was taken, thoroughly mobilized, and used to cover the entire anterior cranial base. Miss Sossia had a remarkably benign post-operative course, complaining of nothing (which may have been related to the disruption of her frontal lobe). I am very sad to report that our friend Cassio, President-Elect of the International Society of Craniofacial Surgery, died unexpectedly on September 2, 2005, of complications from pulmonary fibrosis.

In 1983, the Chinese government expressed an interest in having someone come to China to teach them about craniofacial surgery, and I was asked if I would like to go. I said, sure, and wouldn't they like to have Kawamoto as well. They agreed, so the expedition of myself, 5 Kawamotos (Lance, Mark, and Michelle also came), and two very large and heavy suitcases of instruments left Hong Kong for Shanghai. There we found T. S. Chang to be a wonderful host, and did a good bit of major craniofacial surgery. I can recall doing a Le Fort III, a hypertelorism, and a hemiarrhinia, and Henry and I each did half of a Treacher Collins while the other lectured. Henry did a monobloc advancement, and when we saw the patient on rounds a day or so later, it looked like a small faucet behind his nose had been turned on I said something like, "Geez Henry, what's going on?", and he replied, "Don't worry, they all do that..."

Henry is fond of showing a picture taken with Dr. Chang and a number of our patients in front of their Institute, and asking which of the faces shown is the most abnormal? It is obviously the one on the far left!

There were other trips to distant places, such as Beirut, at a meeting organized by Nabil Hokayim, who had been a Tessier assistant, and Teheran, where Hamid Dirakshani, a very entrepreneurial fellow who had also spent some time with Tessier in Paris, invited us to speak. (I also operated, somewhat reluctantly, on a man with a large vascular malformation of the lip and tongue; Henry managed to stay out of the OR). Tessier was well known in Iran, and had operated there both before and after the down-fall of the Shah, and has some incredible results in the reconstruction of combat injuries from the Iran-Iraq war. The Baron-Tessier flap was often used, mandibles and maxillas were constructed from cranial bone, and osteointegrated implants were placed by Jean Francois Tulasne. (Jean Francois also placed the implants in Tessier's maxillary bone grafts; one iliac and two cranial bone graftings had been carried out by Tulasne, all under local anesthesia.)

Over the years, Henry's career and mine continued on similar trajectories. Both of us had clinical faculty appointments and were actively involved in teaching residents. Both of us began craniofacial fellowships. Our practices were similar in composition, with a mix of craniofacial, orbital, orthognathic, cleft, and cosmetic patients. We both served as President

of the ASMS. I invited Henry to give the Annual Millard Address in Miami, and he invited me to be the Tessier professor at UCLA. We both ended up seeing many of the same patients with often bizarre complaints who were shopping around for Mr. Right as a surgeon. Henry kayaks, I swim.

On a more cordial note, Henry served as the best man at the wedding of Deirdre Marshall and I at Ventana, Big Sur, California, on May 29, 1993.

Craniofacial surgery has changed over these past 30 years. We have better fixation devices, better imaging, and we have distraction osteogenesis. But all of the techniques we learned from Tessier are still valid, and his results on cases done in the 1970s still are better than the best that we can do today. And it was not just surgical technique that we learned from Tessier: we learned how to examine a patient carefully, look for small details, and to diagnose the anatomic problem. In surgery, we learned to have adequate exposure, to be bold and to do as much as possible at one operation, and particularly to avoid all artificial materials and use only fresh autogenous bone grafts. This basic principle of craniofacial surgery seems to be slipping away from some of the younger surgeons in our specialty.

Sometime in the mid 1990s, Tessier took Henry, Kathy, Deirdre, and I on a personal tour of his beloved Bretagne. At the time he was in his late seventies, but insisted on doing all of the driving of our rented Audi, which he did at frighteningly high speeds after many a vinuous lunch. Tessier kept careful notes of his previous visits to the restaurants that we ate at, and these notes, describing, categorizing, and rating what he and Mireille had, were as logical, methodical, thorough, and complete as his patient records.

We continue to work with Tessier on a number of projects. He is in his late 80s now, but as bright as ever, and continues to be a very good source of advice on how to deal with a difficult problem, based on his enormous experience and penetrating intelligence. One of our collaborative efforts, the "T and T" (Tools and Techniques for the Harvesting of Aurogenous Bone Grafts) appeared as a supplement to *Plastic and Reconstructive Surgery* in October of 2005. There are a number of other things that I hope we can finish—The Baron–Tessier flap (clavicular platysmal island), the Arrhinias (Tessier had 51 cases), and a rewriting of the Tessier Classification of Facial Clefts, with a parallel classification of the ageneses (Treacher–Collins–Franceschetti syndrome, for example, moves from being a 6-7-8-cleft to an agenesis; CAAC: Classification of Clefts and Ageneses).

Recently I have heard Henry make noises about retiring, but he should just stop doing that: he is doing his very best work now, and should recall that Tessier continued to do excellent work until he was close to (and even beyond) 80. Henry has trained many excellent fellows, and he owes it to the specialty to carry on and train a few dozen more.

1 Honing the Edge

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INTRODUCTION

Initially, the fellows wanted me to write about the misadventures we shared during their training. Although much can be learned from reviewing unfavorable events, just to jot the incidences down would consume several tomes. In its place I have chosen a topic more manageable, appropriately applicable, and hopefully of equal reading interest—the training of fellows.

Learning craniofacial surgery extends well beyond identifying the malformations, memorizing the steps of an operation and taking care of each patient's special needs. The fellows, all posing superior intellectual curiosity, quickly realize these basic requirements.

However, mastering the *craft* of surgery requires a different set of skills. Old habits must be shed and new disciplines infused.

Apologies to Abraham Lincoln, but all are *not* created equal. The “gifted” are few in any profession and even those who are gifted seek coaching and fine tuning by others. Rarer still is the surgeon whose surgical skills cannot be improved, efficiency enhanced, and time in the operating room reduced considerably. Refinement is a career-long process.

Unfortunately, too much of surgical education is based on the “See one, do one, and teach one” principle. This method is not only inefficient but obviously a little hazardous for the patient. It would be far better to lay the basic foundations at the very start of surgical training with a course entitled Craft of Surgery 101—The Mechanics of Cutting and Sewing. Regrettably, such a course is a rarity.

Once the basic skill sets are ingrained, the next logical step should be directed toward the efficient use of these dexterities. Honing this surgical edge ought to be a part of any surgical residency and certainly of a fellowship.

Why, regardless of task, do some move quickly and effortlessly while others prod along at a sloth-like pace? A surgeon who moves at such a slow, glacial speed might explain that it takes time to do things right; nonsense! A needless minute of anesthesia and time wasted in the operating room never improves a patient's well being.

What then are the defining differences between amateur and professional? Once identified, how should they be articulated and taught? Reflecting upon these issues led me to three seemingly unrelated experiences I have had. The influences that these experiences had on me follows.

MENTORS, RACING, AND TIME AND MOTION

Mentors

The importance of our teachers cannot be denied. I was blessed with mentors who were brilliant surgeons and wonderful people. The talents to teach and motivate are given. Admirably, in addition, they had the ability to laugh at their own foibles and were fun to be with when not troubled.

During training, didactic lectures tell of disease and deformity, treatment plans and surgical expectations. Indication meetings enlighten us of approaches and timing. M&M

(morbidity and mortality) conferences steer us away from pitfalls and teach us judgment. Regrettably, learning the technical skills of operating is left to observation and acquisition by osmosis.

One valued detail cannot be learned from mentors. As I was assisting Dr. John Marquis Converse and Paul L. Tessier (Figs. 1 and 2), in a naïve way I would think, “Technically I can do an operation in a similar manner, but what made those surgeons stand out as great?” I, like a novice chess player, was pondering the next move. They, as masters, were viewing the end of the game and even the result of several operations in the future.

Racing

Many weekends while attending medical school were spent at racetracks—not the type where bets are waged, but where sport cars go fast (Fig. 3).

There are plenty of parallels between race car driving and operating: performing quickly but safely, walking the course before the race to allow study of the ups and downs, camber of the road, sharpness of the curves, roadside markers that could be used as brake points, escape routes, etc. (“the anatomy”). Practice runs gave hints of the very last brake point that could be used, the proper “line” in and out of curves, where to compensate for “over steer” or “under steer,” and how to gain maximum revolutions per minute (rpm) before hitting the straight away (the fellowship).

Other than competitive sports car racing, no experience in my life has come close to the *sustained focus* it demanded. Survival depended upon mental preparation for the unexpected. Once the flag dropped, every bit of attention is highly concentrated into one goal—finishing as fast as possible and safely. The check offs learned during the practice runs are compressed in time. No doubt it was a far better way to spend 30 to 60 minutes than lying on a psychiatrist couch.

It would be ideal to perform operations in this manner. It should be our goal. Unfortunately, it does not occur in the post fellowship “academic” world and rarely in the “real” world.

My first teaching job was at the Sport Car Club of America racecourse. Then, like now, it was instructing without pay. Similarly, the rule was never to be the passenger in your student’s car. This is very much like the adage of keeping sharp things out of the resident’s hands.



FIGURE 1 John Marquis Converse, M.D., Chief of Plastic Surgery at Institute of Reconstructive Plastic Surgery, New York University, autographing his book to Cassio Raposo do Amaral, M.D., Campinas, Brazil, 1979.



FIGURE 2 Paul L. Tessier, M.D. reading a newspaper in Musée de la Pêche, Concarneau, France, 1997. S. Anthony Wolfe posing as mermaid and Henry K. Kawamoto as diver.

Time and Motion

During the later years of dental school, a new educational experiment was conducted on some of us students. The instructors, headed by Dr. Rex Ingram (Fig. 4), called it time and motion, whose principles were long adopted by industry. We jokingly call it time and *commotion*.



FIGURE 3 My weekend medical school assignment: Del Mar Racing track, on to a first-place finish, circa 1960.



FIGURE 4 Rex “Silver Fox” Ingraham, D.D.S., legendary head of restorative dentistry at University of Southern California School of Dentistry who oversaw the Time and Motion program. He is still magnificently sharp and fit at 91 years of age.

In reality, it was a superb exercise. If all surgeons followed its principles, they would save *years* of time during their career!

What was it? Simply put, it scrutinized the waste of time. The squander was enormous. Each dental procedure (operation) can be broken down into components of instruments used and movements of the body required to complete each task. If the number of instruments and their passage between the dentist and assistant can be reduced, the time consumed by the operation will automatically be shortened. If the body is ideally positioned, movements will be more efficient and less fatiguing.

The concepts are simple. But have they ever been taught to surgeons?

The Honing

So, how is it all put together? Like anything else, it begins with proper *prerequisites*. Then comes the *preparation*, followed by the *implementation*.

Prerequisites (Motivation, Anatomy, and Operational Knowledge)

Motivation

By the time advanced surgical training is sought, the pyramidal system has narrowed the field in all dimensions. The surgical sets of skills are well established; some good, some bad and some better than others. Regardless of the level of talent, the most important asset that I have always found present is the desire to improve.

For some, flowing hand-eye coordination is innate. It must be groomed in others. Motivation can narrow the gap and make any operation appear seamless. The key is to be efficient and use motion economically. In addition, one must be confident in what he is doing.

Anatomy

From a technical point of view, nothing instills more confidence than a thorough knowledge of anatomy. I am not referring to the basic anatomy that was gleaned over during traditional medical education and is not even taught in the “modern” curriculum.

As an intern about to perform my very first operation, I was expected and had to draw out the anatomy of an inguinal hernia. As the operation proceeded, the blood that was not in the

pictures of the surgical atlas added the dangerous combination of confusion to excitement. Fortunately for the patient, the attending surgeon guided the operation well. As a trainee, this preoperative exercise left a lasting impression and reminded me of walking around the track before a race to capture the nuances of the turf.

It is the command of the ultra fine surgical anatomy that distinguishes surgeons. Possession of sophisticated knowledge of the surgical terrain permits quicker, safer, and more confident passage.

Conversely, ignorance of anatomy forms the base of surgical fear and misadventures.

Operational Knowledge—Steps of Operations

Every operation is composed of a series of logical steps. When well planned, their numbers are kept to a minimum. Rare is the need to change their order. Yet, too often surgeons get stuck on a step for seeming no reason; worst yet they stumble and bumble a few steps backwards! Excellent surgeons do not get distracted; they side step pitfalls or effortlessly modify the plan to suit the occasion and always keep moving forward.

For the 1975 Congress of the International Society of Plastic Surgery, Paul Tessier was assigned the task of demonstrating a Le Fort III operation over a closed circuit live telecast. He reduced the operation down to a number of steps. In later years he showed me his plan, which included approximately 270 steps! Furthermore, to finish in the allotted time he had each step timed!

Of course, this type detailed listing is not necessary for every day procedures. For complex operations such as the separation of the Guatemalan craniopagus twins (2002), I posted a list of the steps, instruments and supplies that would be needed. Every person in the operating room knew what came next, especially during the wee hours of the morning when the actual reconstruction was begun.

Preparation

Each step of an operation requires specific instruments and supplies. Their number should be distilled to the bare essentials needed to get the job done. An excellent exercise for any surgeon is to select the instruments and lay them out in the order of their use for the scrub nurse. Improved immediately will be the flow of the operation and dramatic reduction in the time spent.

Fatigue should be minimized. It begins by properly positioning the patient and the members of the surgical team. The surgeon's body ought to be situated comfortably to minimize movements. The operative site should be at the elbow height of the surgeon and whenever possible they should sit. Traditionally, dentists stood while working. Time and motion studies eliminated this bad habit and bettered their health.

Uniform illumination of the surgical field reduces visual fatigue. A non-glare finish of the instruments is also helpful.

Implementation—Execution

Of paramount importance are the concepts of time and motion. Wasted movement must be eliminated to reduce fatigue and squandered time. Movement should always be forward and with purpose. How is this done? It has already begun by the orderly placement of the instruments on the scrub nurse's table.

The initial incision tells much about the surgeon. Tentative indecisive surgeons graze the skin with the scalpel, like a chicken who scratches the ground. One should at least get down to the dermis with the first stroke of the "skin" incision; otherwise, it would be called an "epithelial" score. Get down to where the action is. As an example, for a coronal incision, the initial slice should be made directly down to the periosteum or bone.

When an instrument is passed, it is the duty of the surgeon to use it to fruition. That is, complete the job for which it has been designed and with the thought that it will be retired from the operation. This eliminates needless, time frittering, back and forth passes of the same instrument. The same can be said for steps in an operation. Each should be concluded so that it need not be revisited.

An instrument is not like a conductor's baton to be waved around in air. Each instrument should be used with supportive finger, hand, forearm, or elbow rests. In dentistry, they are always used with the little finger steadied on a stationary platform. Using "rests" enhances accuracy, precision, and control.

Keep your eyes on the surgical field. It helps maintain attention. Furthermore, it reduces visual fatigue by eliminating the need to focus and refocus in differently lighted fields.

Give your surgical assistant more work. Their activities should not be confined to holding retractor and cutting sutures. If they are positioned best to do certain portions of the operation, they should. Wasted motion and fatigue thus are further reduced.

Lastly, as in racing, proper pacing is important to preserve energy and heighten efficiency. Activities such as suturing should be automatic and accomplished quickly and effortlessly. Other circumstances call for highly focused attention and patience. A detailed knowledge of anatomy and pathology are important contributors to make this happen. They allow safe passage through difficult terrain where extra attention is required.

SUMMARY

Hopefully, this brief narration will sharpen the habits of surgeons and, more importantly, get better results for their patients. All can improve by following simple principles:

- Refine your knowledge of anatomy and pathology.
- Distill the operation to the smallest number of steps and required instruments.
- Organize self and team.
- Keep mind and eyes focused on the surgical field.
- Increase efficiency and results by reducing wasted time, motion, and frustration.

2 Craniofacial Anatomy

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INTRODUCTION

What can we see when we exam the face of another? What can we feel when we allow our fingers to probe the contours of the cranium? What can we know of pathology by understanding structure? The answer to these questions is “quite a lot.” The experienced craniofacial surgeon can often diagnose a problem in a single glance. What allows the surgeon his diagnostic acumen and his ability to correct the problem is the intimate understanding of the complex three-dimensional structure and function of the craniofacial anatomy.

This chapter is no substitute for a detailed text on head and neck anatomy but rather a focused look at the anatomy through the eyes of a craniofacial surgeon.

ASYMMETRIC FACIAL DISTORTIONS

Symmetry is the hallmark of beauty. The faces of supermodels may vary in ethnicity and coloration but they all share extraordinary degrees of facial symmetry. The human eye can discern minute differences in facial symmetry and it is the first thing that draws ones attention in examination of the craniofacial patient.

Observation of the patient's eyes may yield a great deal of anatomic information. Unequal appearance of the eyes may be the result of a number of differing causes. An eye may appear larger due to unilateral craniosynostosis. In these cases deformation of the greater wing of the sphenoid resulting in a large shallow orbit on the involved side make that eye appear larger. A flattening of the ipsilateral brow, deviation of the root of the nose, and a palpable coronal suture ridge will confirm this diagnosis. A larger appearing eye may be the result of increased orbital contents secondary to tumor or hypertrophy of orbital musculature as in Graves disease. The increase of orbital contents results in *exophthalmos* or the forward positioning of a normal globe in a normal orbit. The normal protrusion of the globe is 14 to 21 mm from the lateral orbital rim to the corneal apex. This is commonly measured with a Hertel exophthalmometer. Greater than 21 mm of protrusion is considered abnormal. We refer to this condition as *proptosis*. As the globe moves forward the upper eyelid position moves upward in its relationship to the limbus. The lower lid will move downward with a resultant increase in scleral show. Inadequate lid coverage of the globe will be demonstrated by an increase in tearing known as epiphora or in severe cases by corneal ulceration. The eye may also appear large due to the orbital volume being too small. A normal orbital content with proptosis is referred to as *exorbitism*. Inward displacement of the orbital walls secondary to trauma or encroachment of the orbital bones as in fibrous dysplasia may result in forward displacement of the globe. A less common reason for proptosis may be a weakness or paralysis of the extra ocular muscles allowing the globe to move forward. In these cases abnormal motion of the globe is also evident. Pulsatile exophthalmos should raise suspicion of an orbital arterial-venous malformation or meningoencephalocele.

The author acknowledges *Fundamentals of Maxillofacial Surgery* (Ferraro J, ed. New York: Springer, 1997) as a major resource for many of the anatomical illustrations in this chapter.

A smaller appearing eye may be the result of congenital *microphthalmus*, inadequate development of the globe itself. Since the growth of the orbit is dependent on the pressure exerted by the developing globe, in these cases the orbit itself is seen to be small. A similar deformity may be seen in children who have been treated with radiation therapy to the orbit. Again inadequate growth of the globe results in a small orbit. Most commonly a smaller appearing eye is the result of an increase in orbital volume usually as a result of prior trauma. The retro position of a normal globe in an enlarged orbit is known as *enophthalmos*. As the globe moves back the upper lid drops in its relation to the limbus a condition known as *pseudoptosis*. The eye may also appear smaller secondary to true ptosis of the upper lid either congenital in nature or secondary to malfunction of the levator muscles.

Orbital *Dystopia* is the uneven position of the orbits in the horizontal or vertical plane. This may be the result of a congenital malformation, childhood tumor, frontal sinus mucocele, or posttraumatic displacement. Vertical dystopia is characterized by the uneven levels of the canthal tendons.

Though symmetric in nature the cranial facial surgeon will also note the distance between the eyes. In general the distance between the eyes is the same as the distance of the palpebral fissure, which is the distance from medial to lateral canthus of any one eye. The normal intercanthal distance is 30 to 35 mm with an interpupillary distance of approximately 55 mm. A widened intercanthal distance is known as *hypertelorism* and a narrow intercanthal distance is known as *hypotelorism*. It is often said, "The face is the mirror to the mind." The embryology of facial development dictates that the frontal nasal area develops in concert with the frontal lobe and midline structures of the brain. Hypotelorism is often the result of inadequate brain development. Associated hypoplasia of the nasal and premaxillary elements is often seen in these cases. Hypertelorism is often the result of a central mass effect separating the orbits during development. This mass may be a frontal encephalocele, tumor, or mucocele. Hypertelorism is also seen in children with central facial clefts and syndromic craniofacial abnormalities.

Critical observation of the patient may reveal other asymmetries, which are a sign of underlying anatomic pathology. Deviation of the root of the nose is seen in association with unilateral cranial synostosis. The root deviates toward the side of the closed suture. Asymmetric ear position is associated with cranial plagiocephaly. In fact anterior displacement of the ear position is the key in differentiating between positional and synostotic *plagiocephaly*. Positional plagiocephaly is characterized by the anterior placement of the ear on the side of the flattened occiput. In craniosynostosis the ear is pulled toward the synostotic suture. The ear is posteriorly and inferiorly displaced in lambdoid synostosis. The height of the normal ear should align the top of the helical rim with the eye brow. Low set ears are often associated with mental retardation in a syndromic child. A small or absent ear, microtia, is associated with hemi facial microsomia characterized by underdevelopment of first and second brachial arch derivatives. In addition to hemi facial microsomia, generalized facial asymmetry may be the result of hemi facial hypertrophy or Romberg's hemi facial atrophy, a unilateral progressive loss of facial soft tissue. The animated face may also demonstrate facial asymmetry as a result of facial nerve paralysis.

FACIAL HARMONY

In addition to facial asymmetries the craniofacial surgeon will be aware of the patient's facial harmony and its relation to the underlying skeletal structure. The concept of facial harmony or the aesthetic balance of the face relies upon the proportion of the major facial structures to each other. From a mathematical standpoint many have observed that the proportion of the facial components may be defined by the *Divine Proportion*, or phi, the Greek letter assigned to the number 1.618... (Fig. 1). This numeric concept defines a line "A" divided into two segments, a longer segment "B," and a shorter segment "C." The ratio of B to C is the same as the ratio of A to B, and this ratio is 1.618. The face abounds with examples of this ratio. If the distance from the eyes to the mouth is A then the eyes to the base of the nose would be B, and the base of the nose to the mouth would be C. If we define A from the chin to the base of the nose then the chin to the

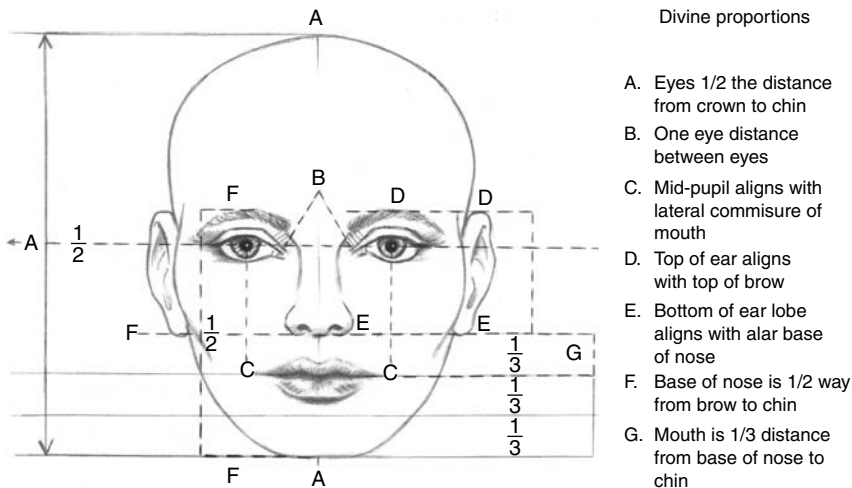


FIGURE 1 Illustration of mathematical depiction for facial proportions. Based on the *Divine Proportion*, or Greek letter phi, assigned to the number 1.618. The mathematical relationship of the distances is $B/A = C/B = 1.618$. *Source:* Illustration provided by Linda Blue.

mouth would be B, and the mouth to the base of the nose would be C. These measurements all fall within the divine proportion of 1.618. There are many more examples of this proportion through out the face.

A less mathematical analysis of facial proportion is that used by portrait artists and is more readily appreciated during a patient office exam (Fig. 2). For the artist the eyes lay half way between the crown of the head and the chin. There is an eye's distance between the eyes and a line dropped from the mid-pupil will align with the lateral commensure of the mouth. The top of the ear aligns with the eyebrow and the bottom of the ear with the base of the nose. The base of the nose lays half way between the eyebrow and the chin. And the mouth lies approximately $\frac{1}{3}$ the distance from the base of the nose to the chin.

Craniofacial surgeons refer to the distance from the nasal radix (skeletal point "N") to the base of the columella (skeletal point "A") as the upper facial height. The distance from the base of the columella to the tip of the chin (*menton*) is referred to as the lower facial height and the upper and lower facial height should be equal. In profile the relationship between the

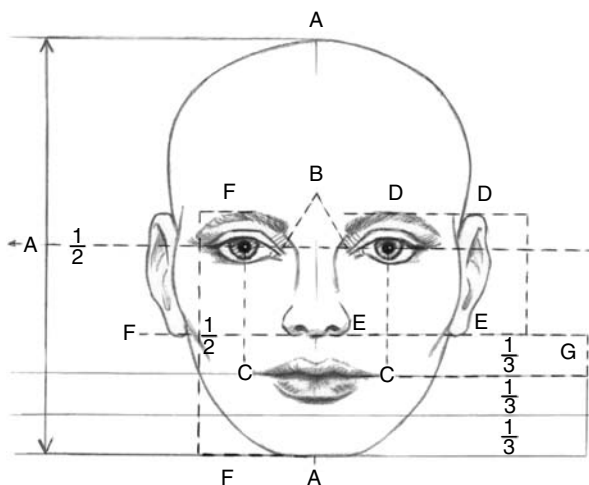


FIGURE 2 Depiction of facial proportions used by portrait artists. This method is much more useful during patient exams and involves a less mathematical analysis. *Source:* Illustration provided by Linda Blue.

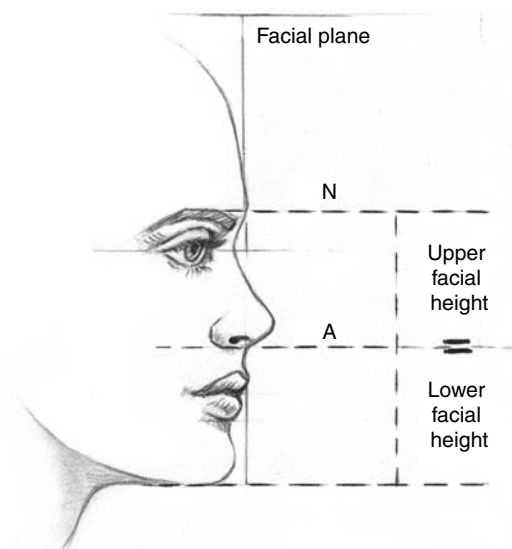


FIGURE 3 Illustration of profile measurements assessed by the surgeon. *Source:* Illustration provided by Linda Blue.

cranial base, the maxilla, and the mandible are described by the *facial plane*. Formal cephalometric studies allow a more mathematical evaluation of these relationships, however, as an easy clinical analysis if one imagines a vertical line to the cranial base dropped from the radix, which is the base of the columella and the chin tip, should all lie upon this line (Fig. 3). A quick assessment of the facial plane during a clinical exam may help to determine whether a patient with an under bite has *prognathism*, an enlarged mandible or maxillary hypoplasia.

CRANIAL DISTORTIONS

There is a wide variation in the shape of the cranium. Familial traits and ethnicity all affect the shape of the head. The Asian skull often shows mid-occipital flattening considered “lucky” by the Chinese, while Masai in East Africa often demonstrate a high conical appearance to their shaven heads. When examining the head of a child it is always helpful to examine the heads of the parents as well. Demonstrating that under their hair the parents may have the same shape as the child often alleviates parental concern for the shape of their child’s head. Distortions of the skull are determined by their origins. In the newborn, normal head size with unusual cranial shape and overlapping sutures is most often due to molding of the skull during birth and does not require X-ray diagnosis or helmet therapy. Molding distortions will generally self correct in the first two months of life. Alternatively the child who presents with craniosynostosis will demonstrate classic patterns of deformity. Patterns of these deformities are due to the inability of the skull to expand in a direction perpendicular to the stenosed suture. An outline of these deformities is presented below (Fig. 4).

- Sagittal Synostosis—*Scaphalocephaly* (keel shaped skull). Long anteroposterior (AP) diameter, bitemporal narrowing, prominent occiput.
- Coronal Synostosis—*Brachycephaly*. Short AP diameter, bitemporal widening, flattened occiput. Bilateral “harlequin” sign on x-ray.
- Unilateral Coronal Synostosis—*Plagiocephaly* (twisted skull). Frontal flattening on affected side, enlarged orbit on affected side, ipsilateral ear pulled toward affected suture, deviation of the root of the nose toward the involved side, unilateral harlequin sign on x-ray.

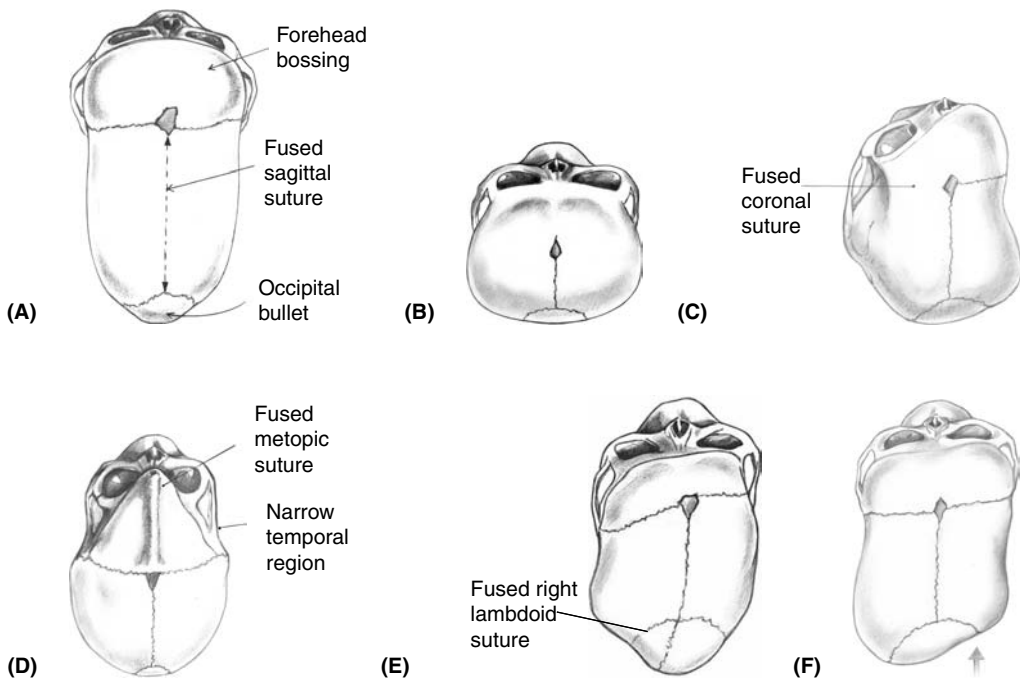


FIGURE 4 An artist's illustrations depicting the six abnormal skull shapes of craniosynostosis. (A) Sagittal synostosis, (B) coronal synostosis, (C) unilateral coronal synostosis, (D) metopic synostosis, (E) lambdoidal synostosis, and (F) positional plagiocephaly. As indicated with a label, the fused suture is shown and the resulting deformity of the skull. Source: Illustrations provided by Linda Blue.

- Metopic Synostosis—*trigonocephaly* (triangle skull). Mid-frontal ridge, frontal temporal narrowing, decrease interorbital distance.
- Lambdoidal Synostosis—occipital *plagiocephaly*. Ipsilateral occipital flattening, posterior positioning of ipsilateral ear, compensatory prominence of contra lateral brow, tilt of cranial base to spinal axis.

Physical examination of the synostotic sutures will demonstrate a palpable ridge over the fused segment.

Positional plagiocephaly is the most common form of cranial distortion one sees today. In 1993 the U.S. Academy of Pediatrics dictated that to decrease the incidence of Sudden Infant Death Syndrome (SIDS), infants should sleep on their back. While the incidence of SIDS has decreased the incidence of positional plagiocephaly has become an epidemic. Infants left on their backs will often prefer turning their head to one side over another. This preference will often lead to flattening of the posterior occiput on the preferred side. *Torticollis*, the unilateral shortening of the sternocleidomastoid muscle will restrict an infant's head motion and may produce a positional plagiocephaly on the involved side.

Positional plagiocephaly is characterized by unilateral occipital flattening, a forward positioning of the ipsilateral ear *away* from the flattened side, and may involve the forward displacement of the ipsilateral brow. In contrast to craniosynostosis, physical examination of the skull will not demonstrate any ridging over the suture area.

In the majority of both craniosynostosis and positional plagiocephaly the head circumference and growth curve remain within normal parameters. Abnormal cranial growth is usually the result of microcephaly secondary to inadequate brain growth or in rare circumstances, pan-synostosis with multiple suture fusions.

EXAMINATION OF THE ORAL AND DENTAL ANATOMY

A pleasant smile is the cross-cultural expression of friendliness and will often succeed where language fails. The degree of tooth show, dental hygiene, and proper occlusion are not only determinants of beauty but also the essential foundation of the facial skeleton. The attractive smile generally shows no more than half of the height of the incisal teeth. A “gummy” smile which exposes the whole tooth and the gum is often the result of vertical maxillary excess or a “long face.” The smile which shows no teeth, when teeth are present, is considered the result of a vertical maxillary deficiency which aesthetically ages the patient and is to be avoided when performing a Le Fort I impaction.

The normal adult has 32 teeth (Fig. 5). Each quadrant of eight teeth possesses a central incisor, lateral incisor, canine, first and second bicuspid, and first, second and third molars. A child’s mouth has 20 teeth. Each quadrant of five teeth possesses a deciduous central incisor, lateral incisor, canine, and a first and second molar. The child’s molars are replaced by the adult bicuspids while the adult molars erupt with out deciduous precursors. Adult incisors possesses a single conical root, the longest being that of the canine. The mandibular molars have two roots while those of the maxilla have three. This is important to note when extracting a maxillary

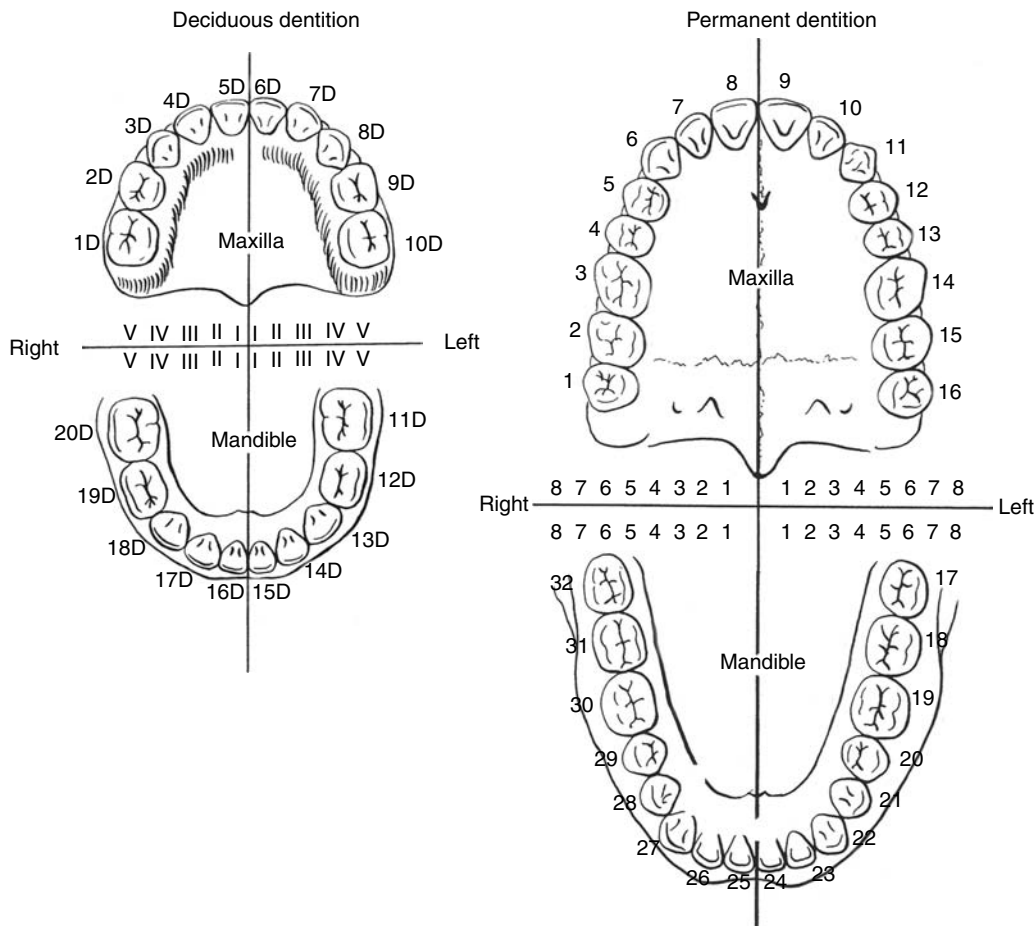


FIGURE 5 Diagrams of dental patterns for children (deciduous) and adults (permanent). Each diagram is divided into four equal quadrants, and numbering of teeth commonly proceeds from the right maxillary third molar to the left maxillary third molar, then proceeds from the left mandibular third molar to the right mandibular third molar. For children, numbering includes 1D–20D, and 1–32 for adults.

molar as one must rotate the tooth buccally to extract the palatal root. One should check the roots of extracted teeth to see that they are complete and that no remnant has been left in the socket.

Numbering of the teeth commonly proceeds from the right maxillary third molar to the left maxillary third molar numbers 1 to 16, then proceeds from the left mandibular third molar to the right mandibular third molar, numbers 17 to 32. Using this system the left maxillary central incisor would be #9 while the right mandibular canine would be #28. The teeth are arranged in an arch form with the maxillary arch being slightly wider than that of the mandible. The maxillary arch displays a slight convex curve from incisor to molars; the mandibular arch mirrors the curve in a convex form. This alignment is known as *the curve of Spee*. Spatial relationships and dental surfaces of the mouth are described as being *mesial*, anterior toward the dental midline. *Distad*, toward the posterior molars, *lingual*, toward the tongue, *buccal*, lateral to the bicuspid and molars, or *labial*, anterior to the canines and incisors (Fig. 6). The occlusal surfaces of the bicuspid and molars are separated into *cusps* and *grooves*, indicating the high and low points of the tooth surface. One refers to the cusp by its position on the tooth as in, the mesial buccal cusp of the first right mandibular molar. Efficient chewing is aided by the cusps of the grinding teeth aligning with the corresponding grooves of the opposing jaw. Molars have four cusps separated by a central groove. Bicuspid by definition have two. *Wear facets* will indicate the actual strike zone of the teeth. For any one individual, the wear facets are as unique as a fingerprint. The presence of *mammalones*, small bumps along the incisal edge of a tooth, indicate that this tooth has never occluded with its partner and hence has never worn down the small irregularities present on a newly erupted tooth. The inability of the incisors to occlude is referred to as *Apertognathia* or an *open bite* deformity. Apertognathia is often the result of premature posterior molar contact secondary to an increase in the height of the posterior maxilla.

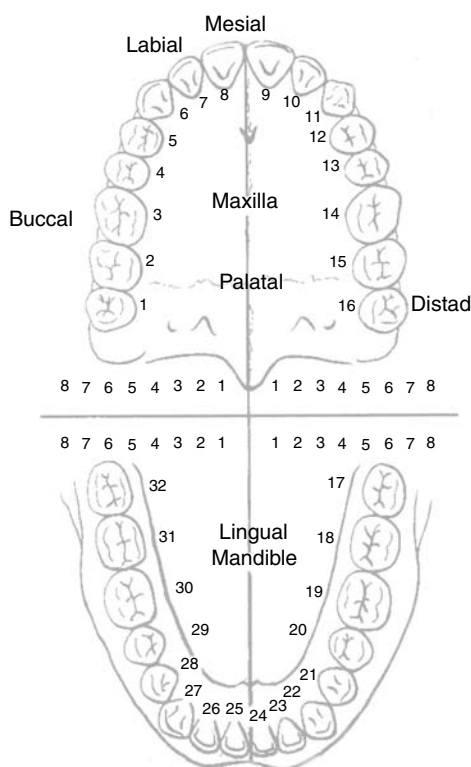


FIGURE 6 The spatial relationships and dental surfaces of the mouth are depicted in this illustration.

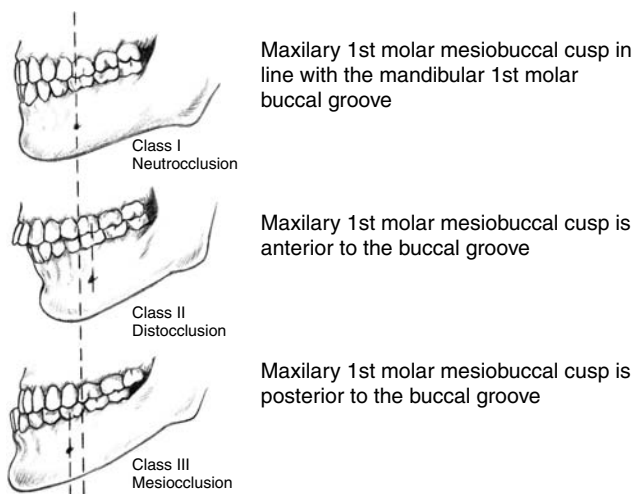


FIGURE 7 The classification system for occlusal relationships, first developed by E. H. Angle, is based on the relation of the maxillary and mandibular first molars to one another. A description and illustration of each of the three different classes is portrayed in this figure.

Dental occlusion is the cornerstone of facial skeletal relationships. In 1899, orthodontist E.H. Angle first classified occlusal relationships based on the relation of the maxillary and mandibular first molars to one another (Fig. 7). A *class I* relationship is defined as *the mesial buccal cusp of the maxillary first molar aligned with the central groove of the mandibular first molar*. This relationship allows for the buccal surface of the maxillary molars to slightly overlap those of the mandible and the maxillary incisors to overlap the incisal surface of the incisors below. The occlusal strike surface of the maxillary incisor is therefore along the lingual surface of the tooth. A small ridge on this surface called the *singulum* is the anatomic strike zone for the mandibular incisor. A *deep bite* relationship exists when the strike zone of the mandibular incisor is high on the neck or gum line of its maxillary partner.

A *class II* occlusion exists when the mesial buccal cusp of the first maxillary molar lies mesial (forward) of the central groove of the tooth below. This produces an overbite situation (Fig. 8). Severe class II relationships occur in Pierre Robin sequence and other conditions which

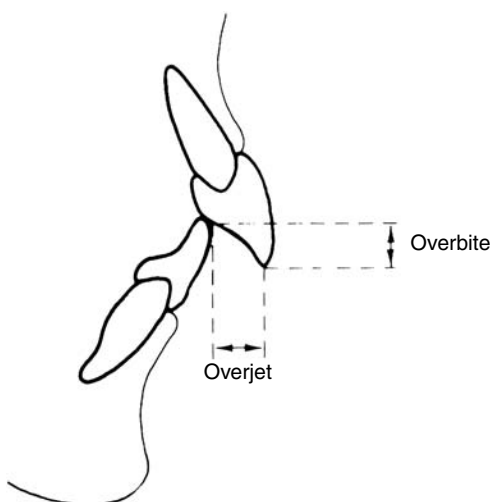


FIGURE 8 Illustration of the overbite, vertical overlap of the incisors, and the overjet, increased horizontal distance between the labial surface of mandibular incisor and lingual surface of corresponding maxillary, which are both associated with class II occlusion.

produce a hypoplastic mandible. We use the term *overbite* to indicate an increased vertical overlap of the incisors. The term *overjet* is used to describe an increase in the normal horizontal distance between the labial surface of the mandibular incisor and the lingual surface of its corresponding maxillary incisor. An increase in the angle of inclination of the maxillary incisor is the usual cause for an overjet relationship.

A *class III* occlusion exists when the mesial buccal cusp of the maxillary first molar lays distad (behind) the central groove of the tooth below. This occlusion produces an under bite. A class III relationship is often observed with the maxillary hypoplasia seen in cleft patients. It may also be a result of mandibular prognathism.

Facial Sensation

The sensory exam of the face may also yield important information on the underlying anatomy. The *trigeminal nerve*, cranial nerve V is responsible for the sensation of the face and scalp (Fig. 9). The nerve is divided into three main divisions arising from the gasserian ganglion, the ophthalmic V_1 , the maxillary V_2 , and the mandibular V_3 (Fig. 10). The ophthalmic division exits through the superior orbital fissure and divides into three major branches, the frontal, the lacrimal, and the nasal ciliary (Fig. 11). The frontal branch exits at the superior rim of the orbit in two terminal branches, the supratrochlear nerve, and the supraorbital nerve. The *supratrochlear* nerve the smaller of the two gives sensation to the conjunctiva, the skin of the upper eyelid, and the medial forehead. In general it

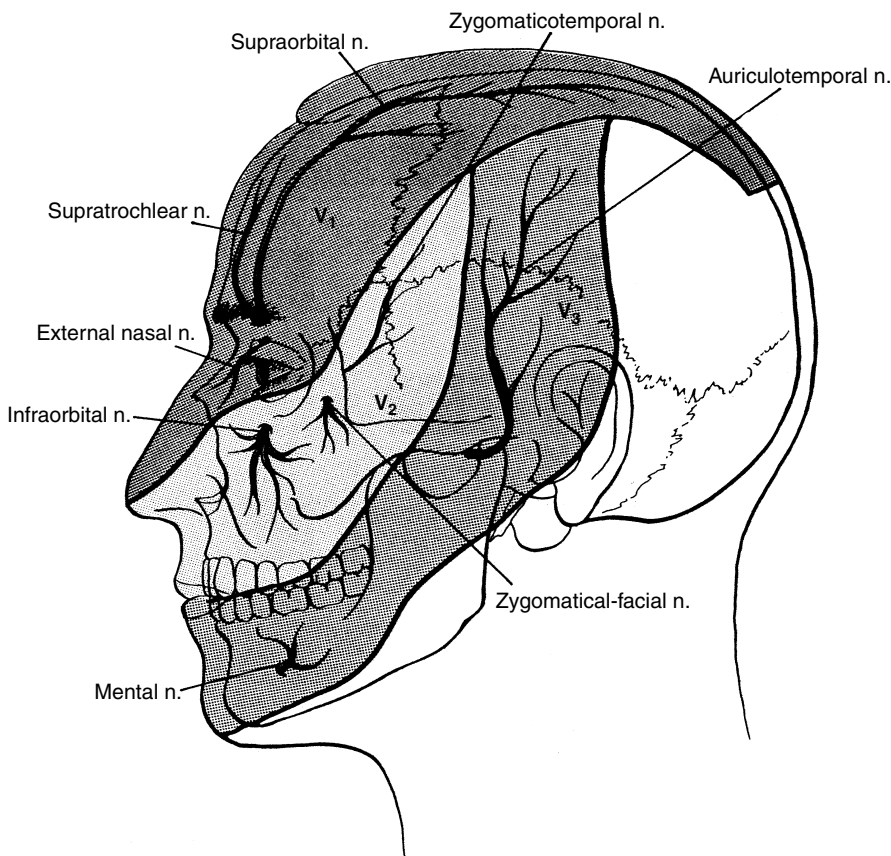


FIGURE 9 Superficial sensory dermatomes from the terminal branches of the trigeminal view.

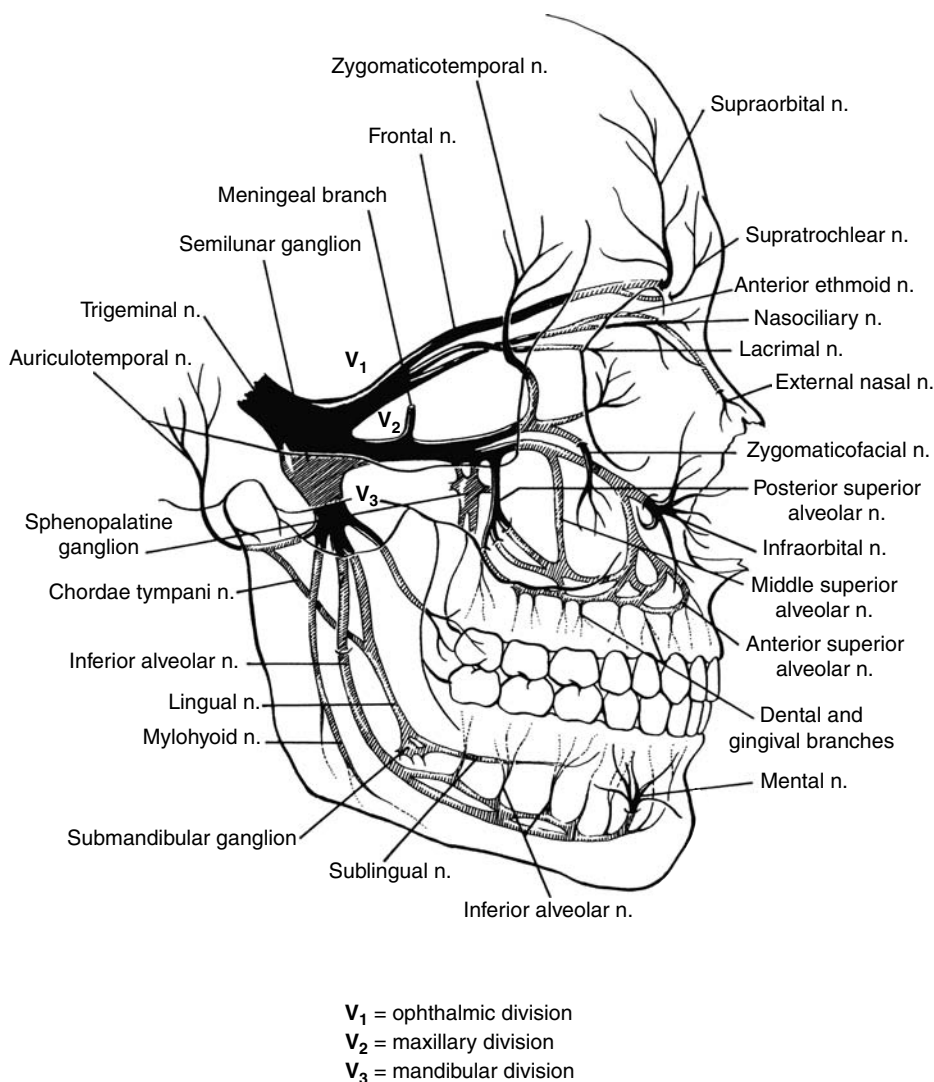


FIGURE 10 The trigeminal nerve (cranial nerve V).

passes medial to the supraorbital nerve and freely under the rim uncontained in a foramen or notch. The *supraorbital* nerve exits the rim at the mid-pupillary line through the supraorbital foramen. The foramen may be complete or exist as an open notch in the orbital rim. A coronal approach to the upper facial skeleton should tease the nerve from its notch or be chiseled from its foramen so that it turns down with the facial flap. Damage to the nerve will result in hypoesthesia of the forehead and scalp as far back as the occiput. The *lacrimal* branch of V₁ provides sensation to the conjunctiva and skin of the upper eyelid. The *nasal ciliary* branch of V₁ carries sensation to the nose, the sclera, and to the cornea of the eye. The nasal branches of the nerve provide sensation to the nasal septum, the lateral wall of the nasal cavity as well as the skin of the ala, and the vestibule of the nose. *Anterior and posterior ethmoidal* branches provide sensation to the frontal and ethmoidal sinuses.

The maxillary division V₂ of the trigeminal nerve exits the skull via the foramen rotundum, courses anteriorly to enter the infraorbital fissure where it runs beneath the floor of the orbit existing via the infraorbital foramen located below the medial aspect of the

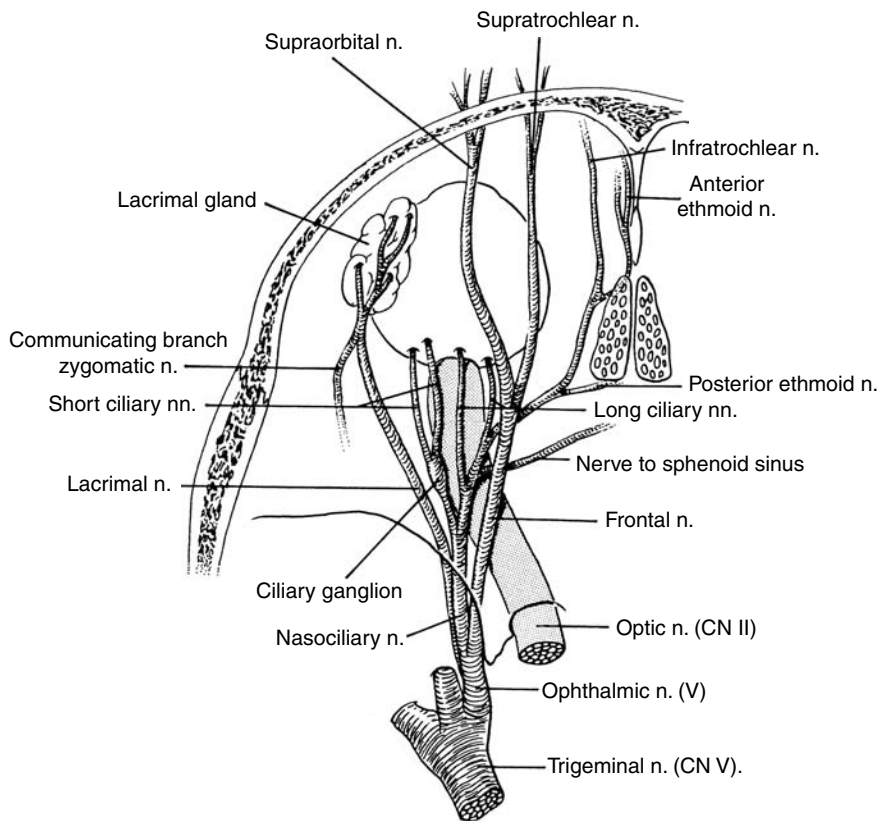


FIGURE 11 Course and branches of the ophthalmic nerve in the orbit.

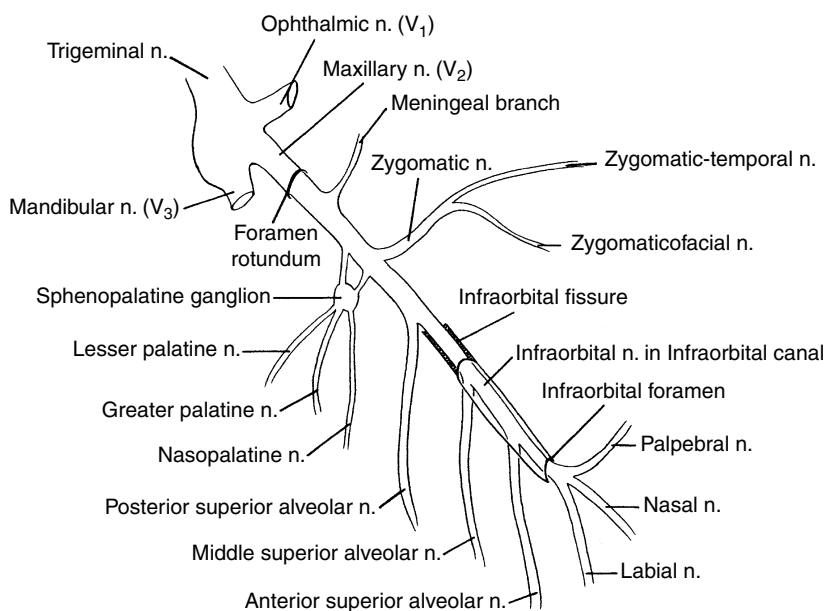


FIGURE 12 Schematic diagram of the maxillary nerve (V₂).

inferior orbital rim (Fig. 12). Along the way the nerve produces several major offshoots. Prior to entering the inferior orbital fissure the nerve sprouts the *lesser palatine*, the *greater palatine* and the *nasopalatine* nerves. The lesser palatine supplies the uvula, the soft palate and the tonsils. The greater palatine supplies the hard palate to the incisive foramen separating the primary from the secondary palate. The nasopalatine runs along the floor of the nose exits through the incisive foramen of the palate and innervates the primary palate along with the alveolar margins of the incisors. Anterior to these branches V_2 produces the posterior, middle, and anterior, *superior alveolar nerves*, the posterior branch supplies sensation to the maxillary first, second, and third molars. The middle branch supplies sensation to the maxillary bicusps and the anterior branch supplies sensation to the maxillary canines and incisors. The terminal branch of the maxillary nerve exits the infraorbital foramen as the *infraorbital nerve*. The infraorbital nerve provides sensation to the skin of the lower eyelid, the anterior cheek, the malar prominence, the anterior temple, the side of the nose, and the upper lip. Because the nerve lies just below the floor of the orbit, orbital floor fractures will often be associated with damage to the infraorbital nerve. Accordingly, patients who present with decreased sensation of the cheek and upper lip after trauma should be suspected of having an orbital floor fracture.

The mandibular division of the trigeminal nerve V_3 exits the cranium via the foramen ovale. The nerve is mixed with both motor and sensory fibers and starts with a main trunk, which then divides into an anterior and posterior division. The nerve primarily functions as the motor supply to the muscles of mastication and sensory supply to the lower jaw and chin. The main trunk of the nerve gives off three motor branches to the lateral pterygoid muscle, the tensor veli palatini, and the tensor tympani. The anterior division of the nerve is primarily motor, and gives off branches to the muscles of mastication in the infratemporal fossa. The *long buccal nerve* is the lone sensory branch of the anterior division and serves to give sensation to the buccal mucosa and lower gums. The posterior division of the nerve gives off the *auriculotemporal nerve* in the infratemporal fossa. The auriculotemporal nerve provides sensation to the temporal mandibular joint, the skin over the temporal region of the ear, the lateral aspect of the scalp, and the tympanic membrane. Further along the division branches the *lingual nerve*, which courses posterior to the retromolar trigone and enters the tongue to provide general sensation to the anterior two-thirds of the tongue as well as the floor of the mouth and the gums. Prior to entering the tongue the lingual nerve receives a contribution from the facial nerve, cranial nerve V_{11} , in the form of the *chorda tympani*. The chorda tympani carries taste fibers to the anterior two-thirds of the tongue as well as autonomic parasympathetic fibers to the sublingual and submandibular glands. The only motor branches of the posterior division are the *mylohyoid nerve* and the *anterior digastric nerve*, which branch just prior to the remaining nerve entering the mandibular foramen. The mandibular foramen lies along the inferior medial aspect of the mandible at the level of the occlusal plane. When V_3 enters the mandibular foramen it is known as the *inferior alveolar nerve*. The inferior alveolar nerve innervates the mandibular teeth by its inferior dental branches as it courses along the alveolar canal within the body of the mandible. The inferior alveolar nerve exits the mandible via the mental foramen and it is then designated as the *mental nerve*. The mental foramen is located roughly between, and inferior to, the first and second bicusps. It should be noted that the mental nerve ascends several millimeters from the canal below to enter the foramen (Fig. 13).

Understanding the relationship of the mental foramen to the canal below it is important if performing a horizontal genioplasty. A cut too close to the foramen may end up entering the canal and damaging the nerve. Damage to the nerve is often associated with mandibular fractures that traverse the inferior alveolar canal. Accordingly, the loss of sensation in lower lip and chin after facial trauma is a high predictor of a mandibular fracture. The inferior alveolar nerve is also put at risk during a sagittal split osteotomy and the surgeon should attempt to hug the lateral cortex of the mandible when performing the osteotomy to decrease the risk of nerve injury.

A multitude of referred pain syndromes may be experienced through the diverse interconnections of the facial nerve. A common example might be pain from dental caries of the mandible perceived through the inferior alveolar nerve, or a carcinoma of the

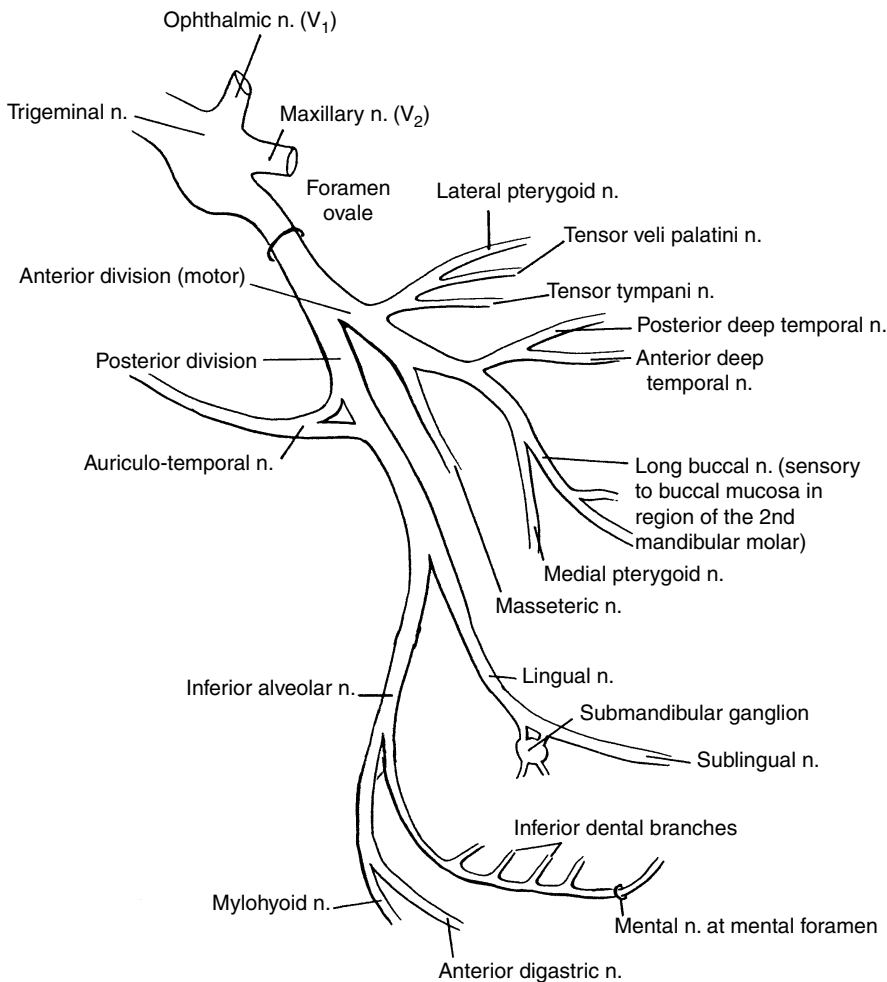


FIGURE 13 Schematic diagram of the mandibular nerve (V₃).

tongue perceived through the lingual nerve, referred through the mandibular branch of the trigeminal nerve to the auriculotemporal nerve and perceived as pain in the tympanic membrane.

The Facial Nerve

The *facial nerve*, cranial nerve V₁₁ innervates the muscles of facial expression. The nerve exits the skull through the stylomastoid foramen, passes under the protection of the mastoid process and courses anteriorly into the substance of the parotid gland where it divides into its five major branches. It should be noted that in a young child the mastoid process is not fully developed and the nerve runs a superficial course unprotected by the mastoid as it passes beneath the ear lobule. A standard facelift incision in a small child puts this nerve at risk for injury.

The five major branches of the facial nerve are the frontal (temporal), zygomatic, buccal, mandibular (ramus mandibularis), and cervical. The *buccal and zygomatic branches* share a multitude of interconnections. As a result function to the associated muscles of the mid-face may be well preserved despite injury to some of these branches. The Frontal and Mandibular

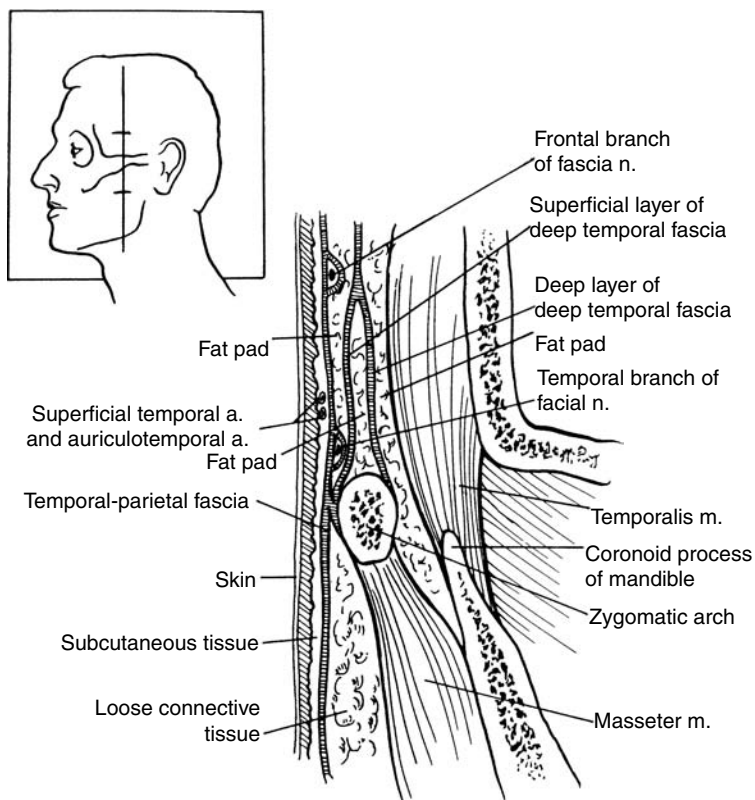


FIGURE 14 Schematic cross-section showing the temporal fascia.

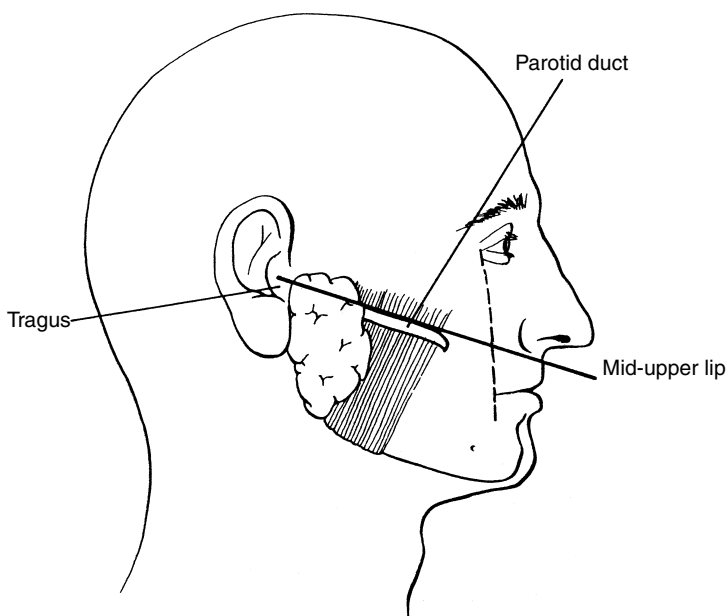


FIGURE 15 The parotid duct is found on a line drawn from the tragus to the mid-upper lip. Anterior to the line drawn from the lateral canthus to the lateral commissure, the facial nerve branches have arborized to an extent where repair is not necessary.

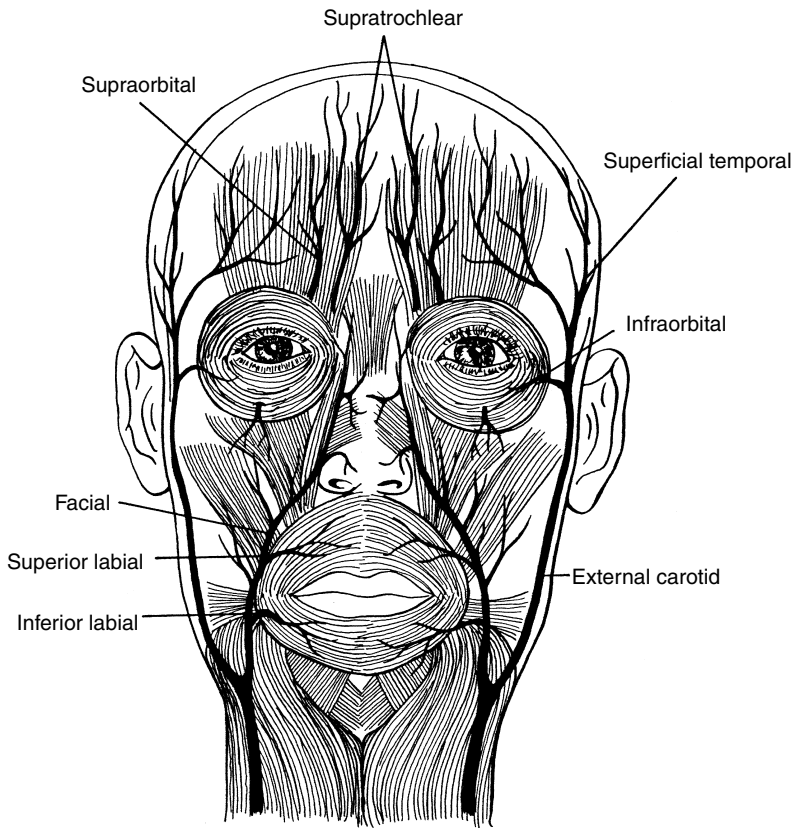


FIGURE 16 Anterior view of facial arteries.

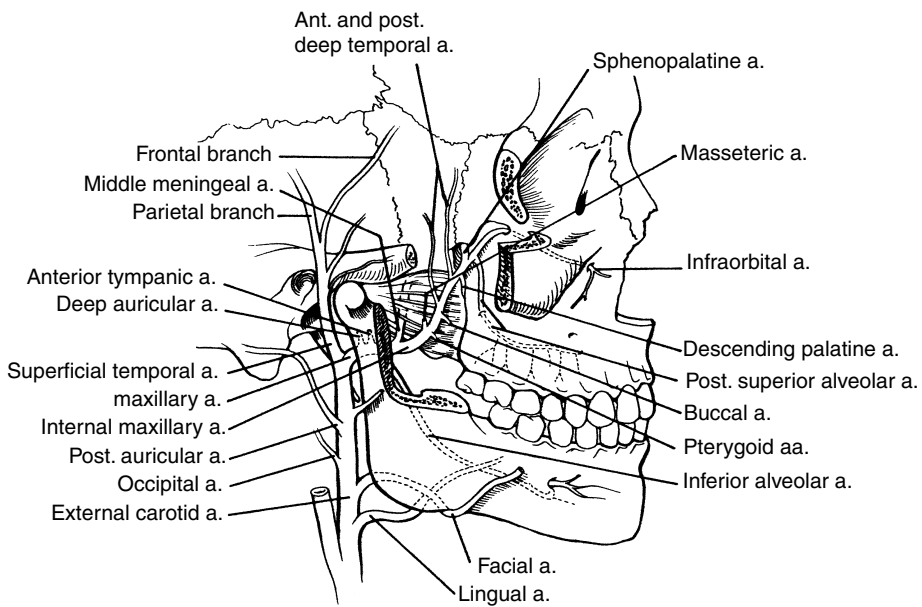


FIGURE 17 The maxillary artery and its branches.

branches of the nerve are more singular in their innervations and damage to these nerves will be immediately evident.

Knowledge of important superficial landmarks is helpful in understanding the underlying course of the facial nerve branches. The *frontal branch* of the nerve crosses the zygomatic arch at a point roughly half way on a line between the inferior helical rim and the lateral cantus. The branch then proceeds to a point approximately 15 mm above the lateral brow. The nerve in its course lies deep to the temporal parietal fascia and above the

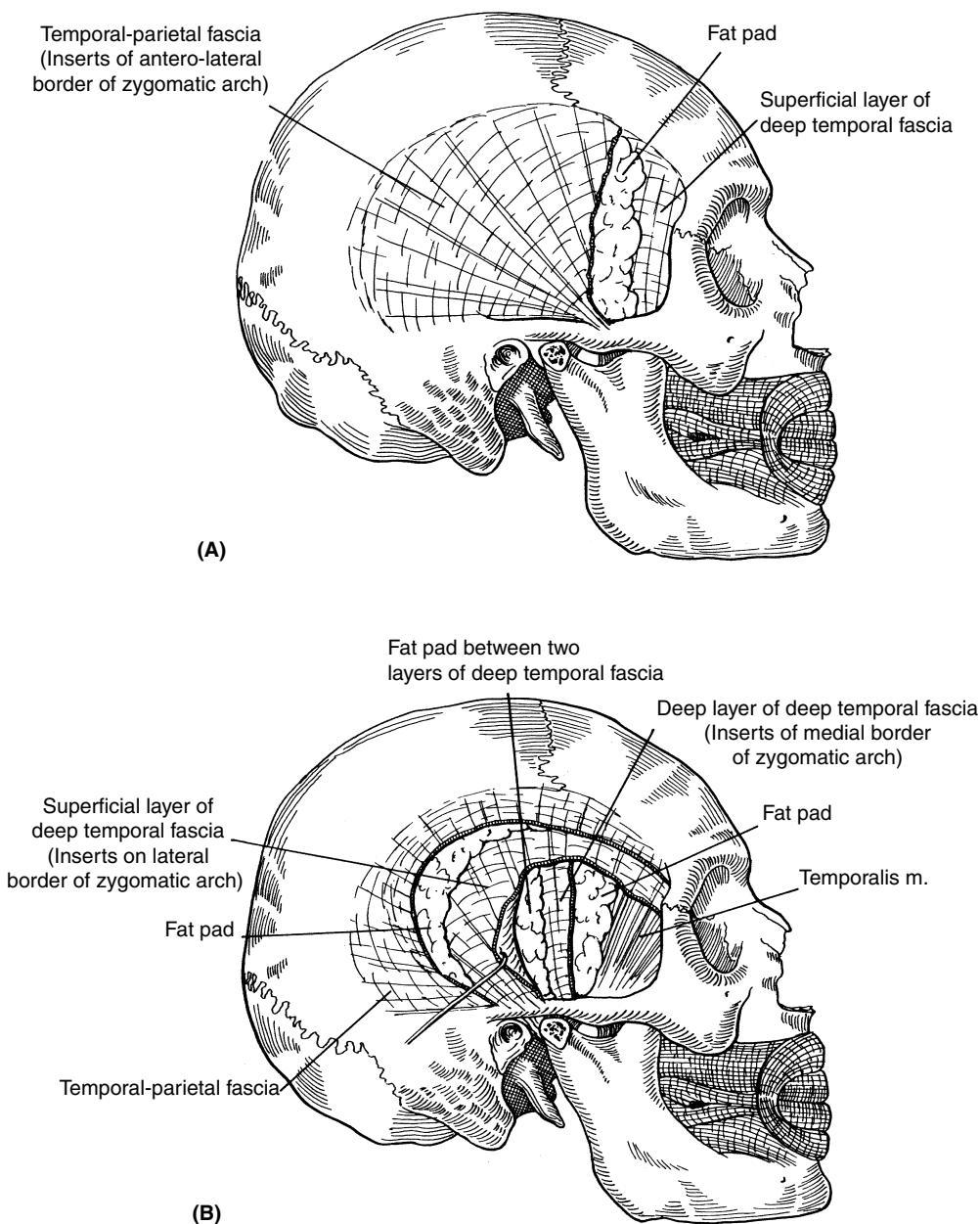


FIGURE 18 (A,B) The temporal parietal fascia, the most superficial layer of the temporal fascia. Cutaway shows fat pad between temporal parietal fascial layer and the superficial layer of the temporal fascia. The deep temporal fasciae, both superficial and deep layers.

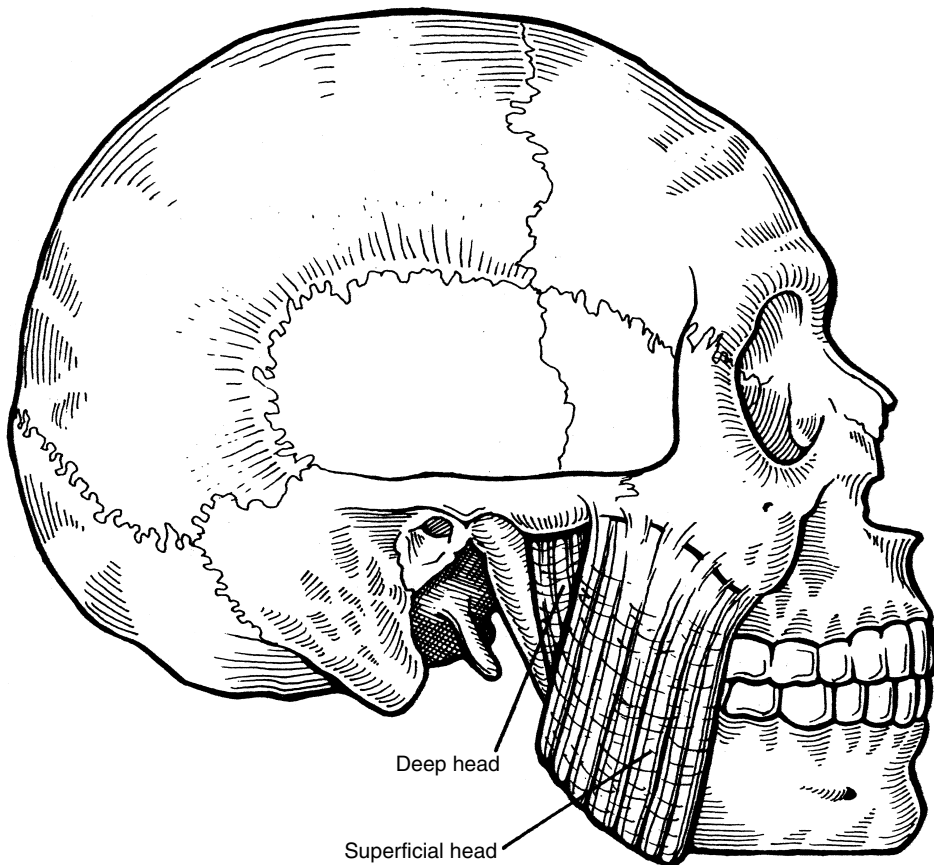


FIGURE 19 The two heads of masseter muscle.

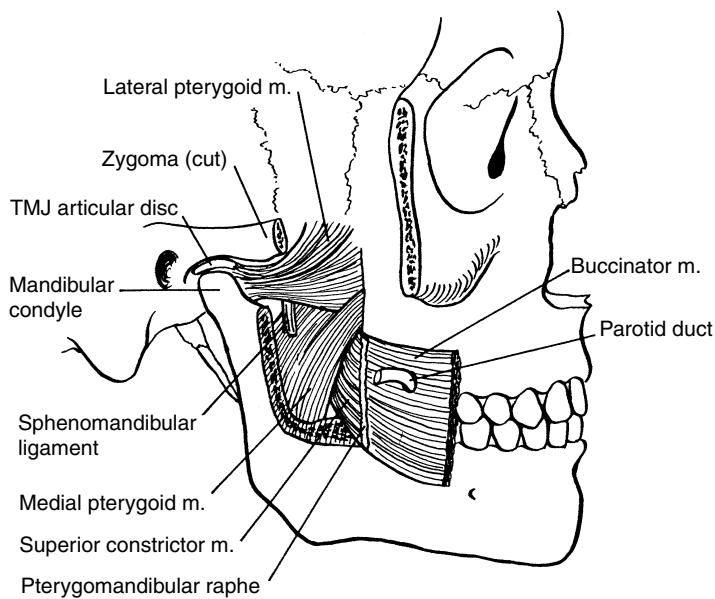


FIGURE 20 The medial and lateral pterygoid lateral. *Abbreviation:* TMJ, temporomandibular joint.

superficial portion of the deep temporal fascia. Coronal approaches to the facial skeleton are best approached at the plane between the superficial and deep portion of the deep temporal fascia to avoid damaging the nerve. Damage to this nerve will present as an inability to raise the ipsilateral brow (Fig. 14). The zygomatic and buccal branches of the facial nerve produce their interconnected branches within the substance of the parotid with multiple arborizations as the nerve courses toward its distal muscular insertions. A line dropped from the lateral canthus of the eye to the lateral commissure of the mouth is a helpful delineation of those arborizations. Anterior to this line the nerve branches are too small to require operative repair. One need not worry about the nerve branches in deep lacerations anterior to this line. Of further note is that the *parotid duct* is no longer present anterior to this line having pierced the buccinator muscle at the level of the anterior boarder of the masseter muscle. The course of the parotid duct lies on along the line described from the tragus to the base of the ala (Fig. 15).

The *mandibular branch* of the facial nerve crosses the inferior boarder of the mandible at the point anterior to masseter muscle identifiable by the palpations of the facial artery. The nerve lies below the platysma and crosses superficial to the artery. On an oblique course from posterior and inferior to the inferior mandibular boarder, to anterior and superior along that boarder. A Risden approach to a mandibular fracture is best performed a centimeter or two inferior to the mandibular boarder retracting the nerve upward and away from the dissection. Damage to the nerve will be apparent by the loss of depressor function of the lower lip on the involved side.

Facial Blood Supply

The facial blood supply is provided by the facial artery and drained by the facial vein. The *facial artery* has its origin on the external carotid arising as the *external maxillary artery* and can be palpated entering the face at the mandibular boarder anterior to the masseter muscle boarder. After crossing the mandibular boarder the artery runs superiorly and medially deep to the platysma, risorius, and zygomaticus. The vessel gives off the inferior and superior *labial arteries* and an alar branch to the base of the ala before terminating in the *angular artery*

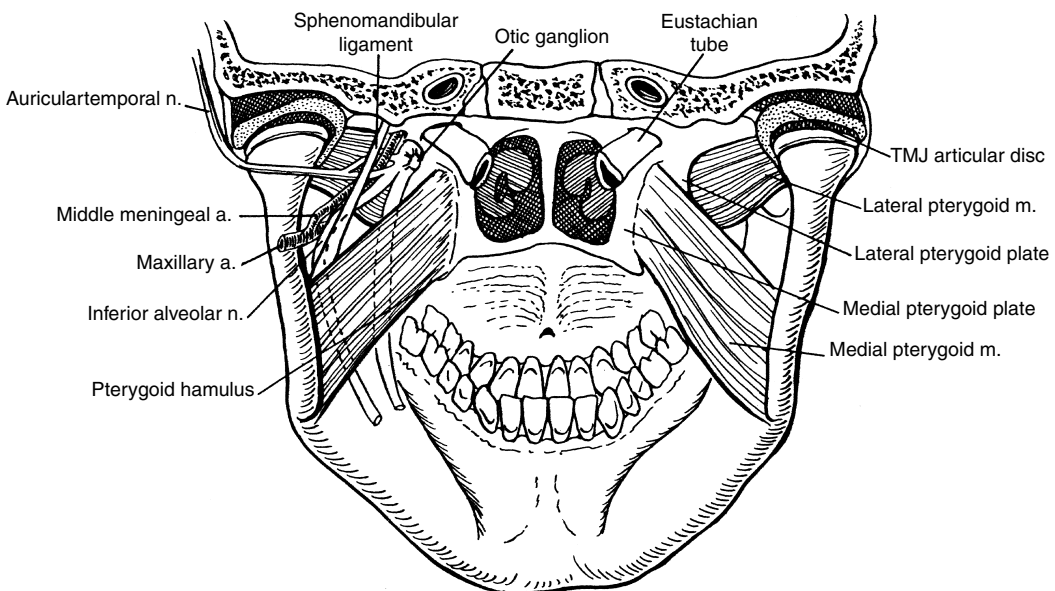


FIGURE 21 Posterior and slightly interior view of pterygoid muscles and their anatomic relations. *Abbreviation:* TMJ, temporomandibular joint.

at the medial angle of the palpebral fissure. The labial arteries run deep to the orbicularis and superficial to the sub mucosa of the lip. The inferior labial artery serves as the axial blood supply of an Abbé flap. The artery's sub mucosal position allows for division of the lip skin and orbicularis muscle with the creation of a thin pedicle promoting an ease of rotation to the flap (Fig. 16).

The *anterior facial vein* courses parallel and posterior to the facial artery, the vein begins as the *angular vein* at the angle of the nose and the eye. The angular vein communicates with the *orbital vein*, which communicates with the cavernous sinus. Infections of the upper

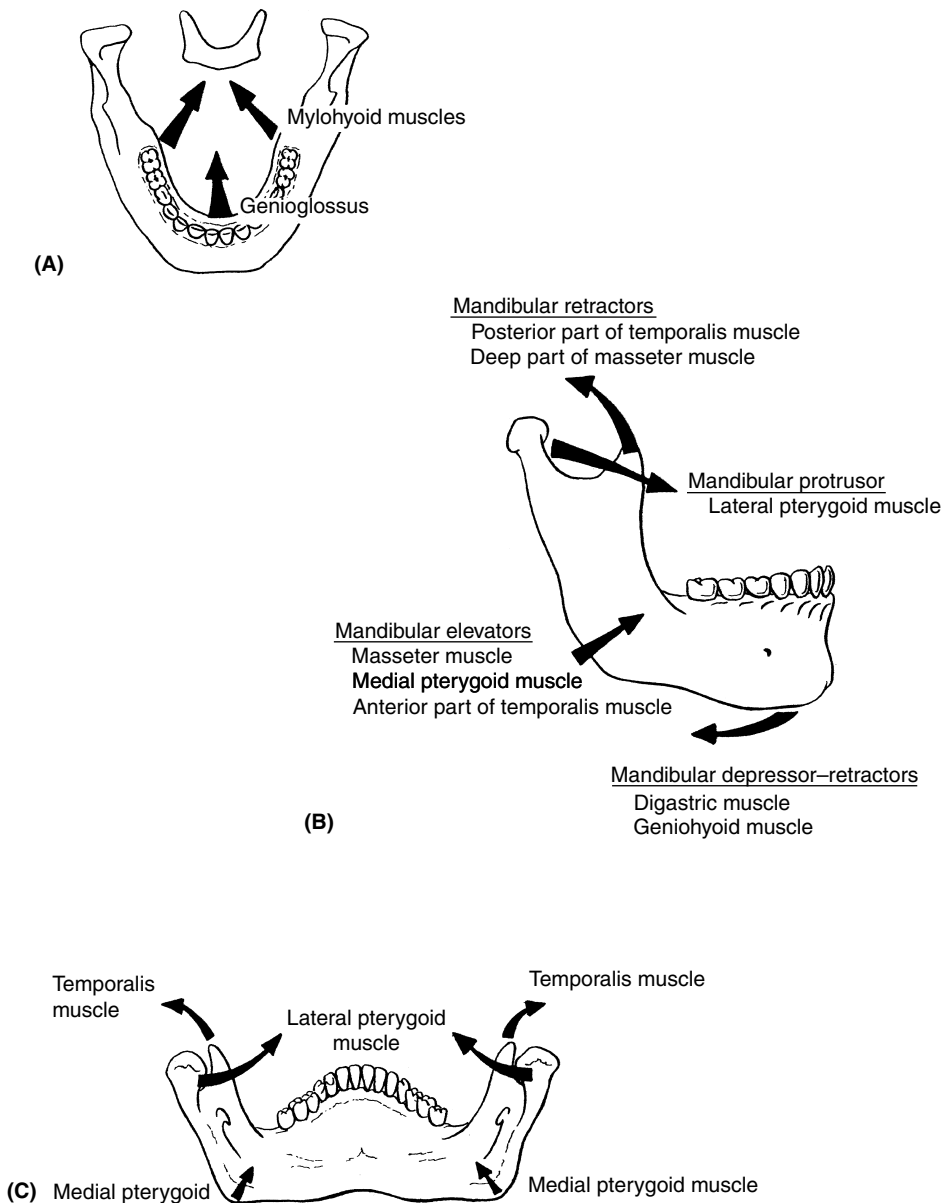


FIGURE 22 Representation of the forces of the mandibular musculature upon the mandible in (A) the anterior muscle group, (B) the posterior muscle group and mandibular depressor-retractors, and (C) the posterior muscle group.

face therefore have the potential for intracranial extension and resultant cavernous sinus thrombosis.

After branching the external maxillary artery, the external carotid divides into the *superficial temporal artery* and *internal maxillary artery*. The superficial temporal artery is palpable anterior to the tragus and subsequently divides into a parietal and frontal branch as it courses in the temporal region. The internal maxillary artery runs in close approximation to the lateral pterygoid muscle in the infratemporal region. The artery branches the *meningeal artery* to the brain, the *inferior alveolar artery* to the mandible and teeth, multiple muscular branches to the muscles of mastication, the *descending palatine* to the palate, the *posterior superior alveolar* to the posterior maxillary teeth, and finally terminates as the *sphenopalatine* and *infraorbital arteries*. The descending palatine artery is in close proximity to the pterygoid plates, and is at risk for injury during the pterygomaxillary disjunction during a Le Fort osteotomy. Keeping the osteotomy low at the pterygoid plates will avoid an increase in blood loss from a severed descending palatine vessel (Fig. 17).

The Mandible and the Muscles of Mastication

The craniofacial surgeon considers occlusion to be the cornerstone of the facial architecture. The mandible is the dynamic foundation for that cornerstone. The mandible articulates with the cranial base at the *temporal mandibular joint* where the condyle of the mandible contacts the glenoid fossa of the temporal bone. The temporal mandibular joint allows for both rotation and forward translocation of the condyle as the mandible swings through its opening arch of rotation. The seating of the condyle in its most posterior unrestrained position is known as *centric relation*. Successful orthognathic surgery is predicated upon the patient being in *centric occlusion*, the teeth in proper occlusion, and centric relation, the condyle being properly seated in the glenoid fossa. The motion of the mandible is provided by the four muscles of mastication the temporalis, the masseter, and the medial and lateral pterygoids. The *temporalis muscle* has a wide origin along the temporal crest of the skull and extends to the superior lateral rim of the orbit (Fig. 18). The majority of the muscle lies in the temporal fossa where it is covered by a superficial and deep temporal fascia. The wide muscle narrows as it passes under the zygomatic arch and attaches to the coronoid process of the mandible. The action of this muscle is that of a rotator and closer of the mandible. The

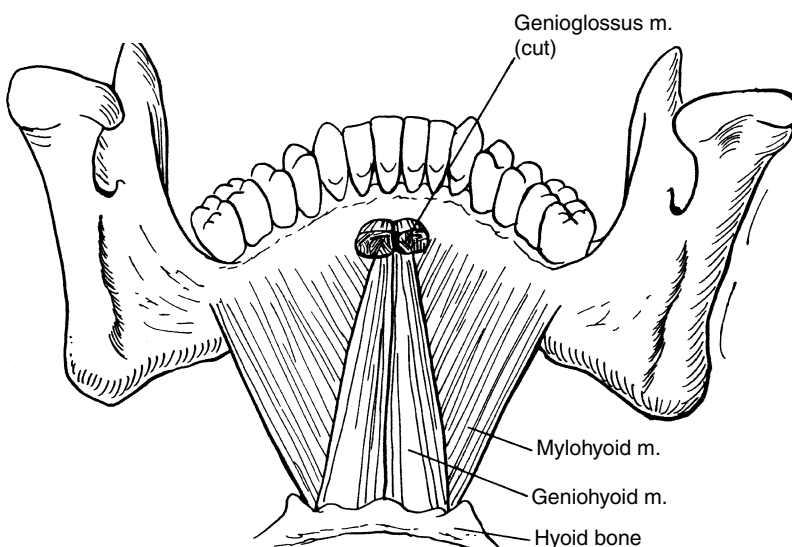


FIGURE 23 Posterior view showing the muscles of the floor of the mouth.

superficial temporal fascia attaches to the anterior portion of the zygomatic arch with the deep fascia attaching right behind it. The plane between the superficial and deep temporal fascia is the proper plane of dissection for a coronal approach to the upper facial skeleton. Reflection of the coronal flap at this level allows the frontal branch of the facial nerve to reflect with the flap, as it is then safely located above the superficial temporal fascia and below the temporal parietal fascia.

A Gilles approach to elevate a zygomatic arch fracture or reduce a malar complex fracture takes advantage of the course of the temporalis muscle. An elevator resting on the muscle in the temporal fossa may then follow the fibers toward their insertion on the coronoid and in doing so will place the elevator below the zygomatic arch and malar complex. An anterior elevation and reduction of the fractures may be accomplished from this position.

In dissecting the upper facial skeleton it is beneficial to maintain the temporalis fibers attaching to the lateral orbital rim. A failure to maintain these fibers during a surgical advancement of the rim may result in a postoperative temporal hollowing.

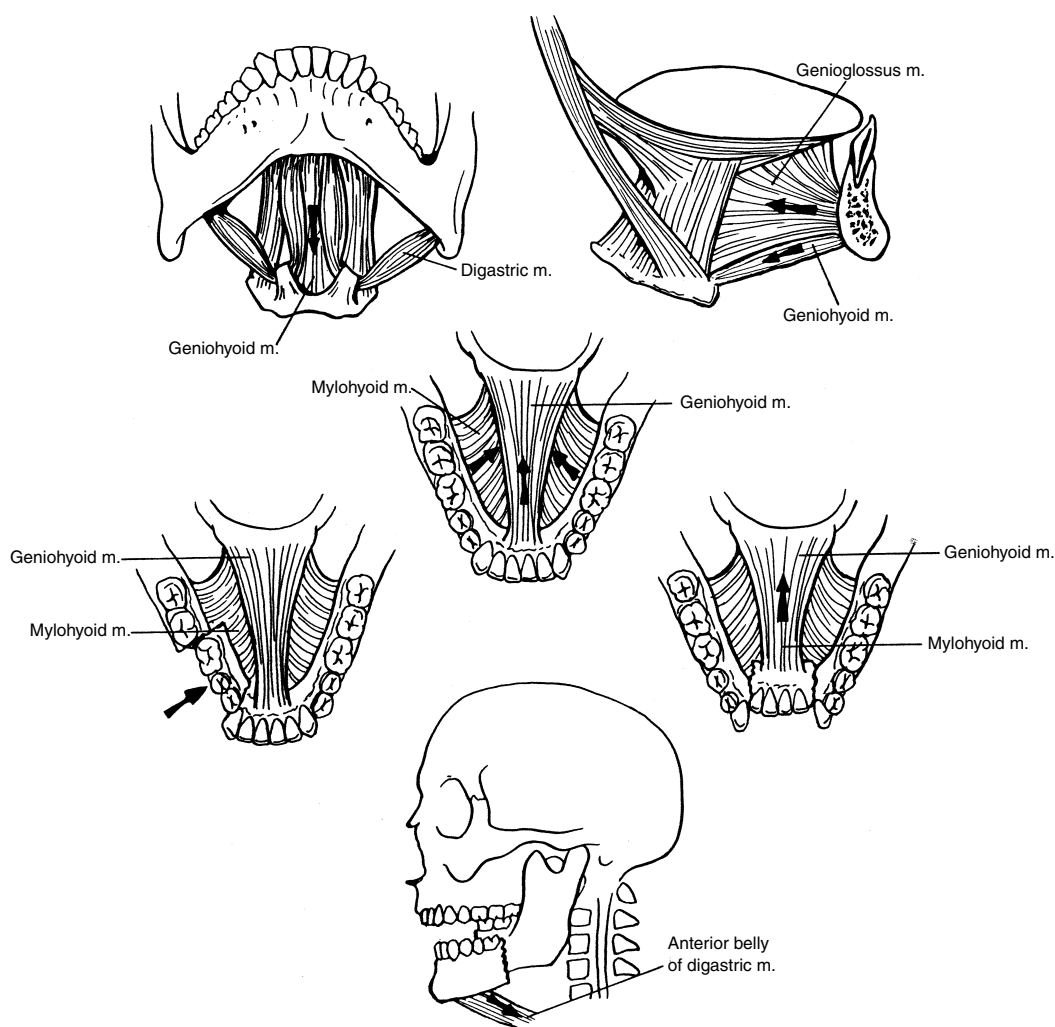


FIGURE 24 Actions of the suprahyoid muscles upon the mandible. Arrows indicate the directional pull of the muscles and displacement of the mandibular fracture fragments.

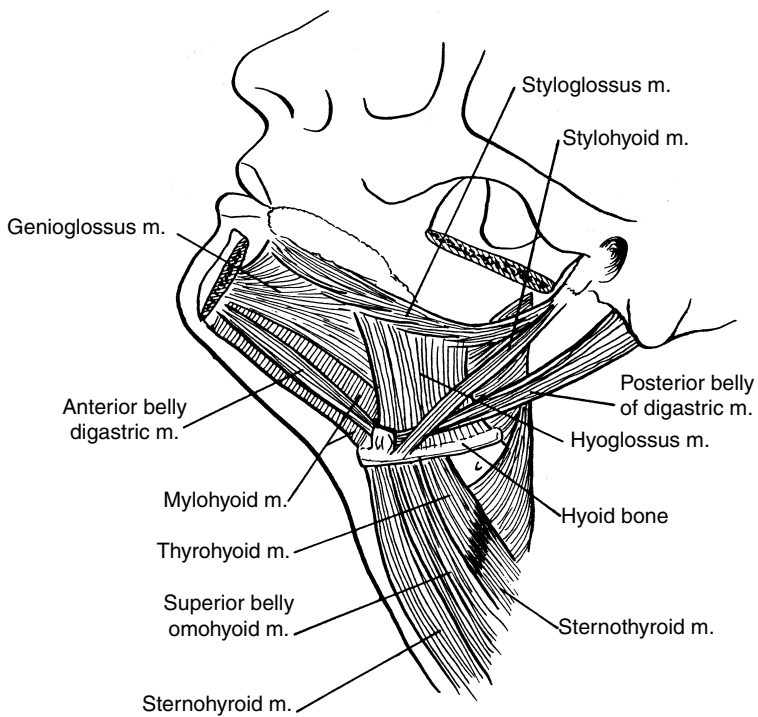


FIGURE 25 The suprahyoid and infrahyoid muscles.

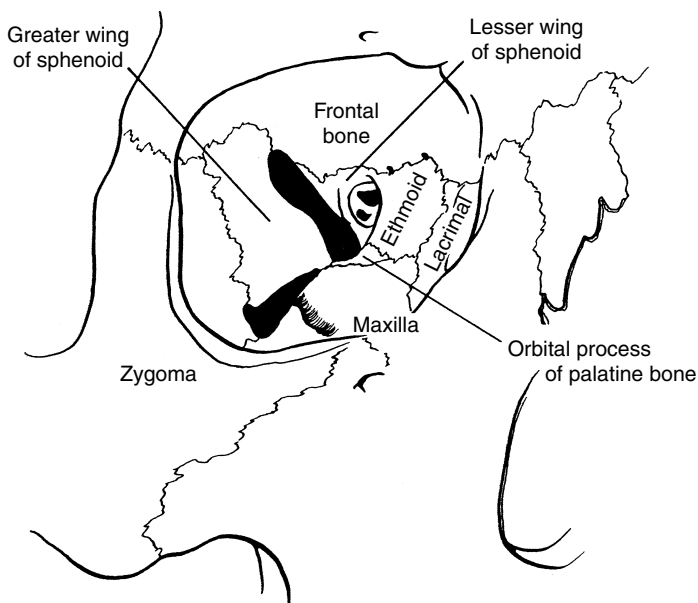


FIGURE 26 The bony orbital anatomy.

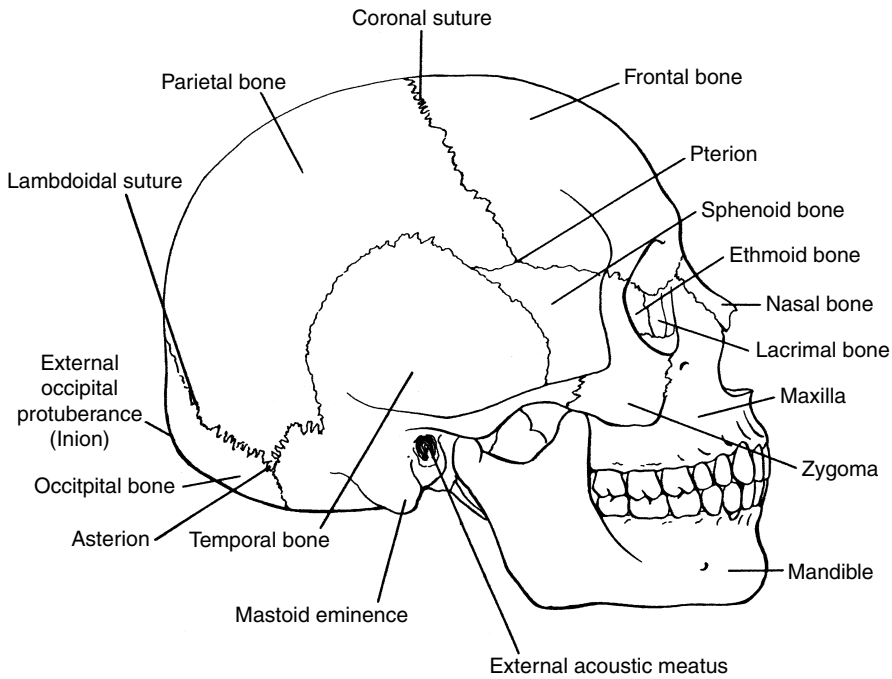


FIGURE 27 Lateral view of the skull.

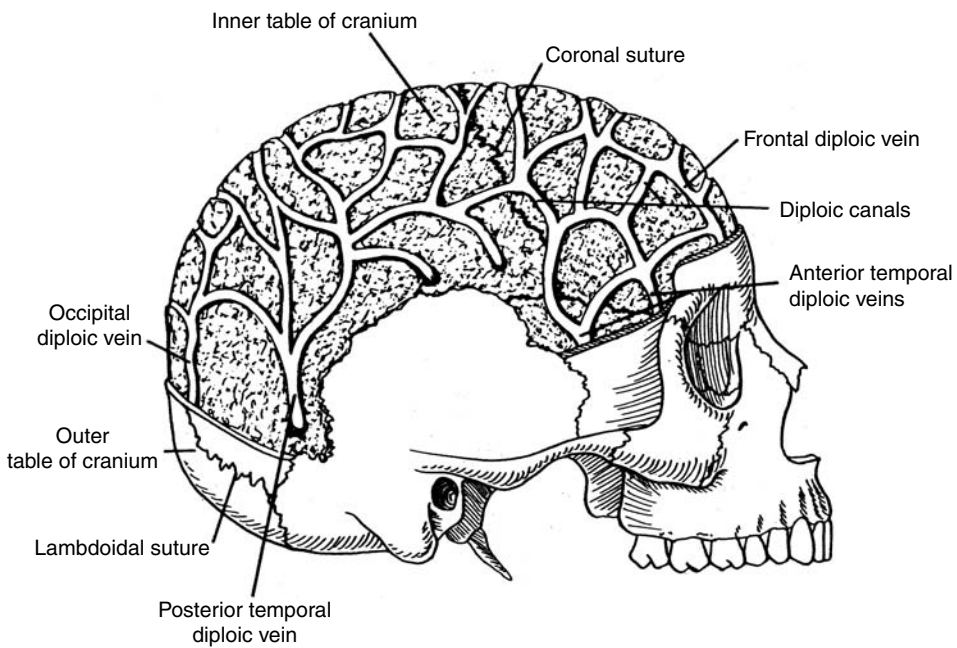


FIGURE 28 The cranial architecture and the four diploic veins.

The *masseter muscle* arises from two heads. The superficial head arises from the lower boarder and anterior two-thirds of the zygomatic arch. The muscle fibers slant inferiorly and posteriorly overlying the deep head of the muscle to insert into the lateral and inferior portion of the ramus of the mandible. The deep head of the muscle originates from the inner surface of the posterior one-third of the zygomatic arch, where it then runs vertically downward to join the insertion of the superficial head of the muscle at the ramus. The masseter muscle functions a powerful closer of the mandible with the deep head also serving as a mandible retractor (Fig. 19).

Though there are two pterygoid plates, the medial, and the lateral, the pterygoid muscles, medial and lateral, both take their origins from the lateral pterygoid plate. The *medial pterygoid muscle* has its origin on the medial aspect of the lateral pterygoid plates. The fibers of this muscle run lateral, inferior, and posterior to insert along the medial and inferior boarder of the ramus of the mandible. In this position the fibers form a sling with the masseter muscle and act as a powerful closer to the mandible.

The *lateral pterygoid muscle* has its origin on the lateral aspect of the lateral pterygoid plate. This muscle has two heads. The superior head of the muscle originates high on the lateral pterygoid plate and the infratemporal surface of the greater wing of the sphenoid. The muscle sweeps posteriorly where it inserts upon the articular surface and disc of the temporal mandibular joint. The inferior head of the lateral pterygoid arises for the lateral surface of the lateral pterygoid plate and proceeds posteriorly where it inserts upon the neck of the condyle at the pterygoid fovea. The lateral pterygoid muscle is a protruder of the mandible. In a Le Fort I fracture the combined actions of the pterygoid muscles may displace the maxillary segment inferiorly and posteriorly, causing premature contact of the posterior molars resulting

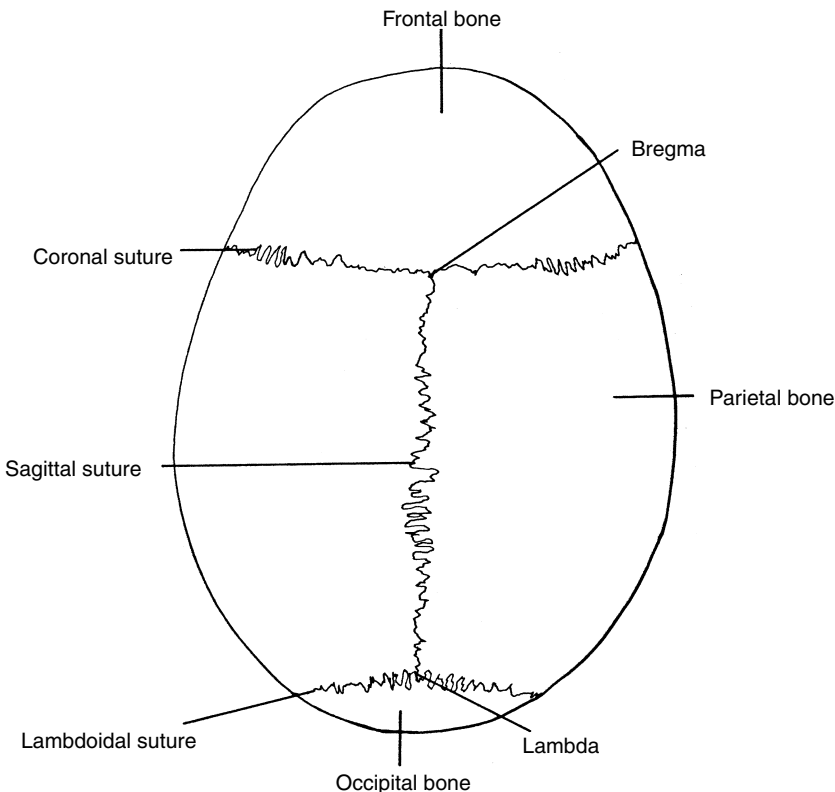


FIGURE 29 Superior aspect of the skull.

in apertognathia, an open bite deformity. The inability to close the anterior dentition is a frequent presenting sign of the maxillofacial trauma patient and can be seen in both Le Fort I fractures and bilateral subcondylar fractures (Figs. 20 and 21).

Though not muscles of mastication, there are several other muscles that attach to the mandible and may exert displacement forces to mandibular fracture segments (Fig. 22).

The *mylohyoid muscle* forms the floor of the oral cavity spanning from the mylohyoid line of the medial surface of one hemi mandible to the other (Figs. 23 and 24). The muscle serves as a tongue elevator during swallowing. In the intact mandible it exerts no effect on the bone. In a fractured mandible it will tend to pull the fracture segments in a lingual direction. Lying on top of the mylohyoid is the *genioglossus muscle* which originates from the mental spine of the lingual surface of the symphysis and extends in a broad fan—shaped manner to insert along the inferior tongue from apex to base. The genioglossus acts as a protruder and depressor of the tongue. In a bilateral parasymphyseal fracture the genioglossus will displace the central mandibular fragment lingually. Lying inferior to the mylohyoid muscle are the mandibular depressors, the digastric, and geniohyoid muscles. The *geniohyoid muscles* are paired muscles whose origin is the mental spine of the inner anterior mandible. The insertion of the muscle is the body of the hyoid bone. The muscle is

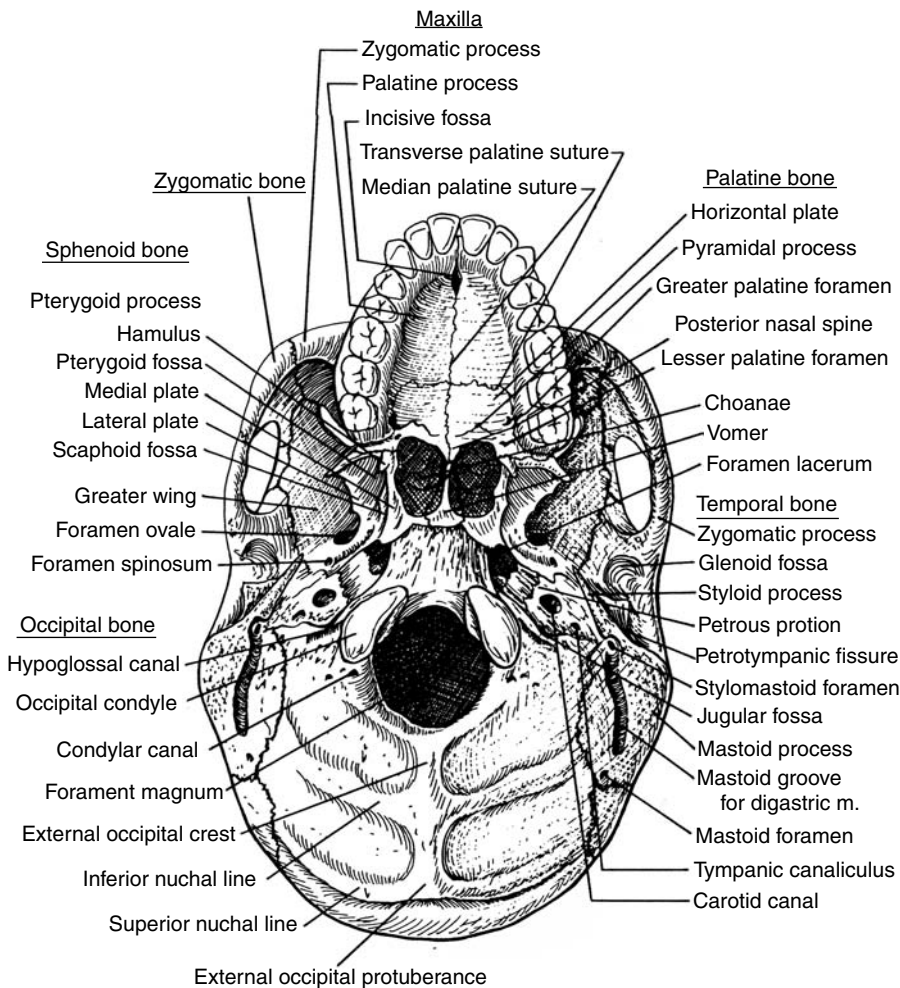


FIGURE 30 Inferior view of the cranial base.

innervated by the hypoglossal nerve (CN XII) as well as fibers from the first cervical nerve. The muscle acts as a depressor and retractor of the mandible. The *digastric muscle* is composed of a posterior and anterior belly connected by a tendon. The posterior belly has its origin on the medial mastoid process. The muscle passes forward and inferiorly becoming tendonous as it passes through a fascial sling at the level of the hyoid bone. The tendon then courses sharply upward and forward becoming the anterior belly of the digastric which inserts into the digastric fossa on the lower medial surface of the mandible. The posterior belly is innervated by the facial nerve (CN V₁₁). The anterior belly is innervated by the mandibular branch trigeminal nerve (CN V₃) that generates the nerve to the digastric and mylohyoid muscles before entering the alveolar foramen of the mandible. The digastric serves as a retractor and depressor of the mandible (Fig. 25).

The Orbit

The bony orbit has the shape of a truncated pyramid. The base of the pyramid is the orbital rim and the apex is most posterior at the entrance of the optic nerve through the orbital foramen. The widest portion of the orbit corresponds to the equator of the globe approximately 1.5 cm from the rim.

The orbit is described as having four sides, a lateral and a medial wall, a roof, and a floor. The floor angles slightly down from the rim at the most anterior portion then slowly gains elevation as it proceeds posteriorly toward the optic foramen. The medial wall lies in the

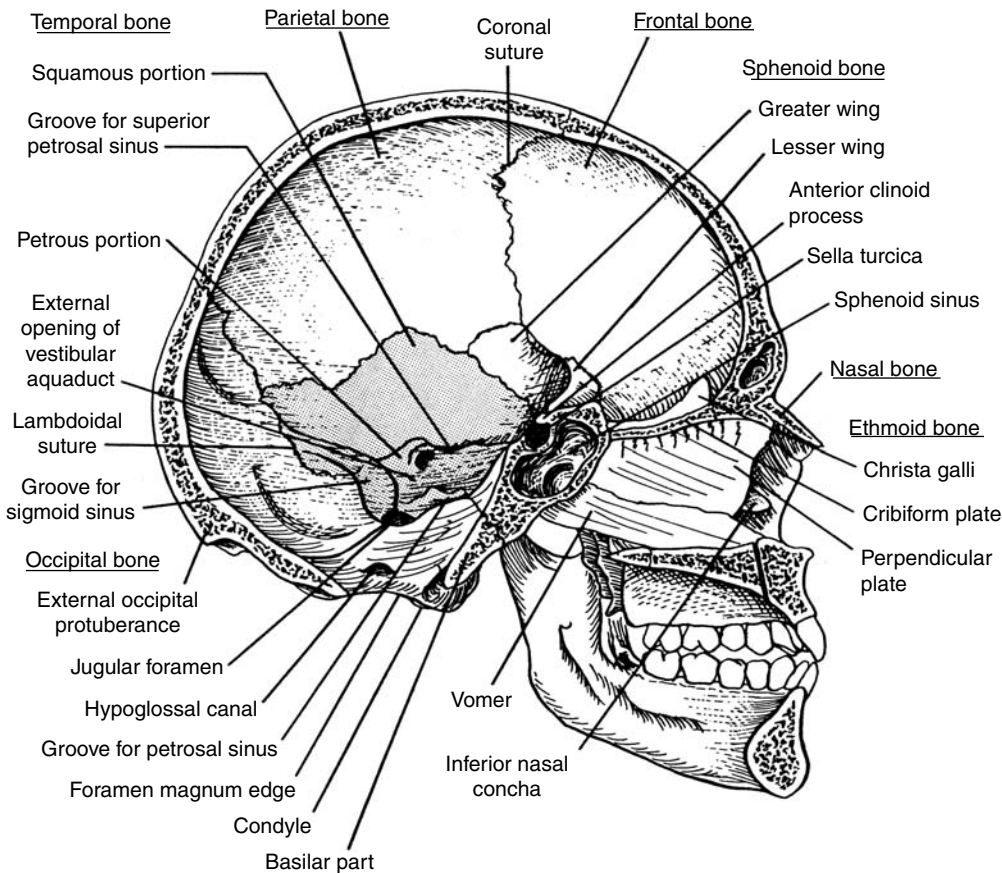


FIGURE 31 Mid-sagittal section of the skull.

sagittal plane of the skull while the lateral wall lies at a 45° angle to this plane placing the apex of the pyramid at the medial aspect of the orbit.

There are eight separate bony portions which form the orbit; frontal, greater wing of the sphenoid, lesser wing of the sphenoid, ethmoid, lacrimal, maxilla, zygoma, and orbital process of the palatine bone. The roof of the orbit is composed of the frontal bone and lesser wing of the sphenoid. The medial wall is composed of the ethmoidal bone and anteriorly the lacrimal bone. The lateral wall is composed of the zygoma and greater wing of the sphenoid, while the floor is made up of the maxilla, comprising the roof of the maxillary sinus, and a small posterior segment of the palatine bone. The continuity of the floor is broken by the *inferior orbital fissure*, which communicates laterally with the infratemporal space lying medial and posterior to the lateral orbital rim. Medially the fissure branches into the *infraorbital canal* which accommodates the infraorbital nerve. The orbital floor is weakest along the canal and therefore orbital floor and malar complex fractures frequently occur along this line, resulting in compression of the infraorbital nerve with resultant hypoesthesia to the anterior cheek region. Orbital blow out fractures often affect the medial wall of the orbit as well. The medial orbital wall separating the orbit from the ethmoidal air cells is known as the *lamina papyracea* (paper wall) and as the name implies, it represents an area of structural weakness. In addition to the *optic foramen* the orbit communicates intracranially via the *superior orbital fissure* running between the greater and lesser wings of the sphenoid. The *oculomotor* (CN III), *trochlear* (CN IV), and *abducens* (CN VI) enter the orbit via this fissure (Fig. 26).

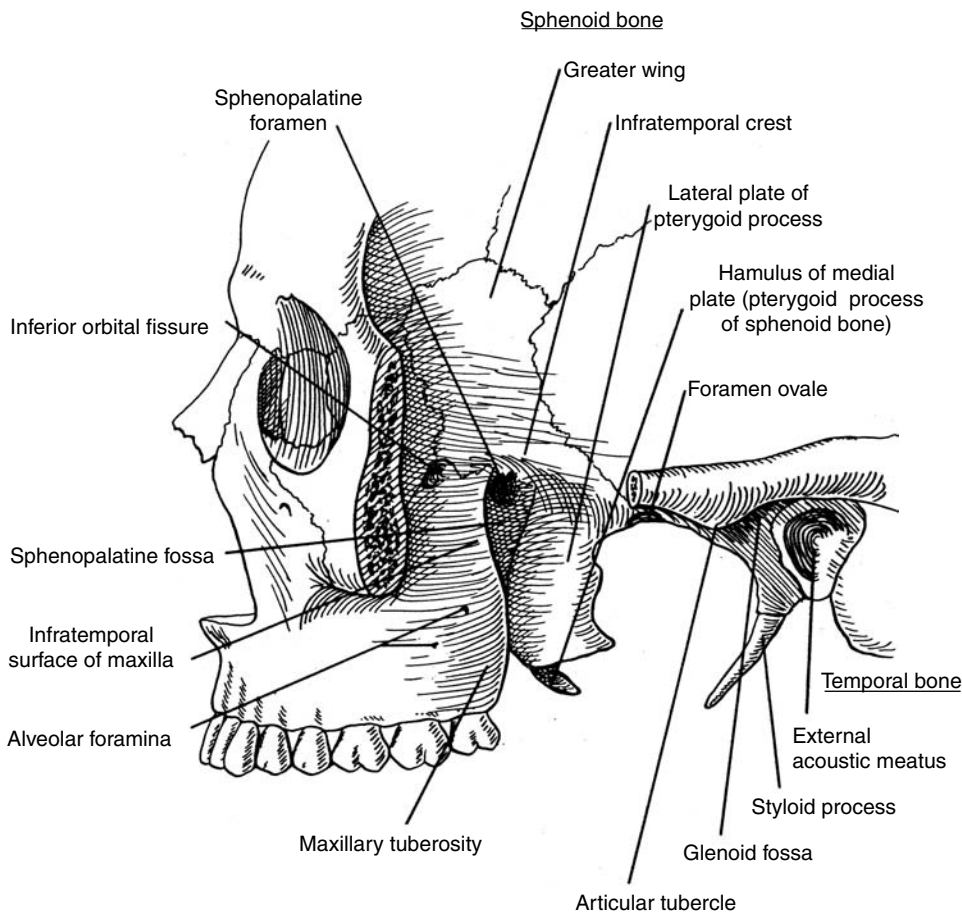


FIGURE 32 Infratemporal fossa exposed with zygomatic arch removed.

Orbital fractures may be approached via atransconjunctival, sub ciliary, or lid crease incision. All of the approaches should then enter the sub periosteal plane of the orbital floor. Dissection within this plane will elevate the orbital contents from the floor. The *optic nerve* lies approximately 44 mm from the orbital rim along the medial wall of the orbit. A helpful landmark to gauge the depth of ones dissection is the posterior wall of the maxillary sinus, approximately 38 mm from the orbital rim and easily palpated with an instrument through the orbital floor fracture. Additional helpful landmarks are the anterior and posterior ethmoidal

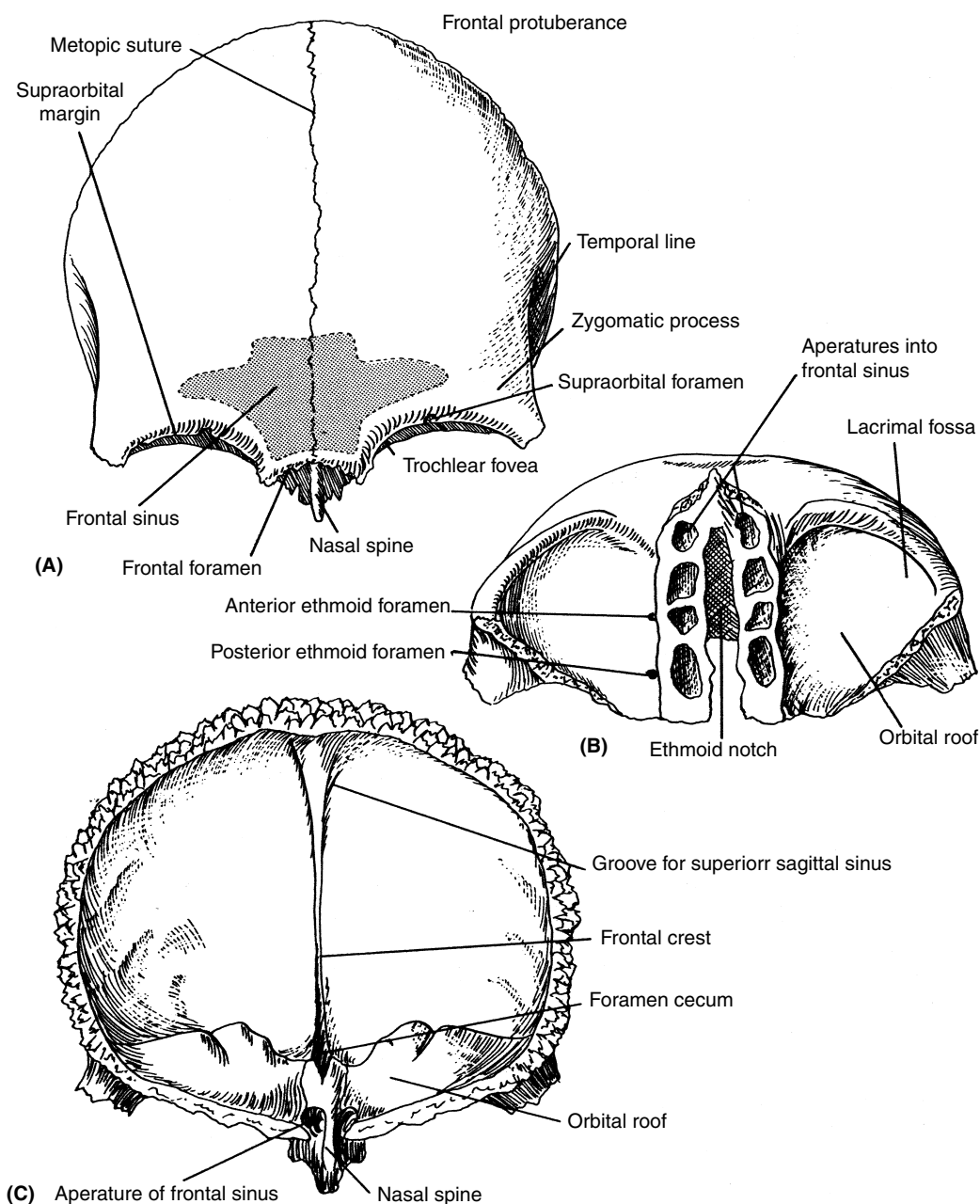


FIGURE 33 The frontal bone from the (A) anterior, (B) inferior, and (C) posterior views.

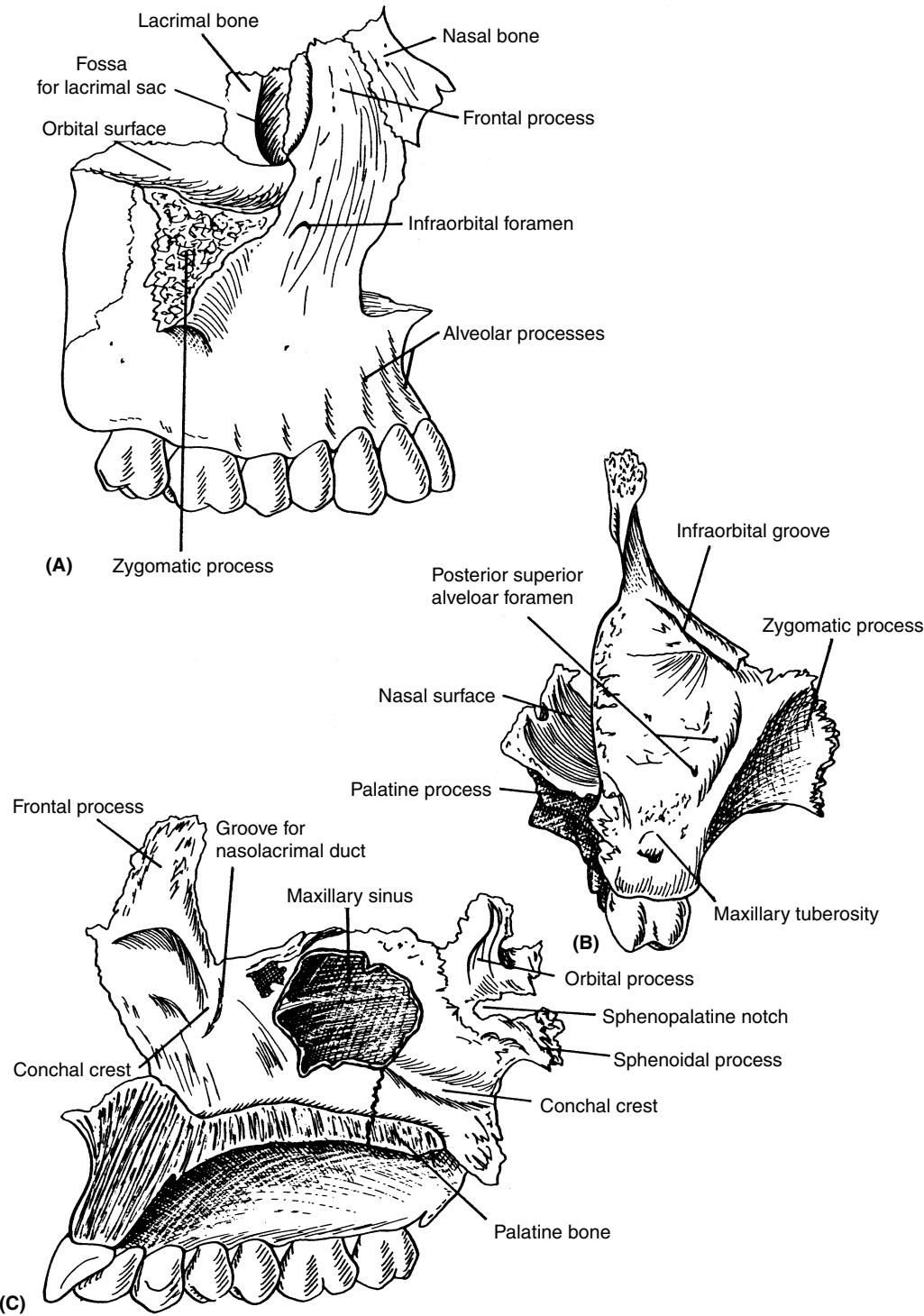


FIGURE 34 The maxilla from (A) lateral, (B) posterior, and (C) medial views.

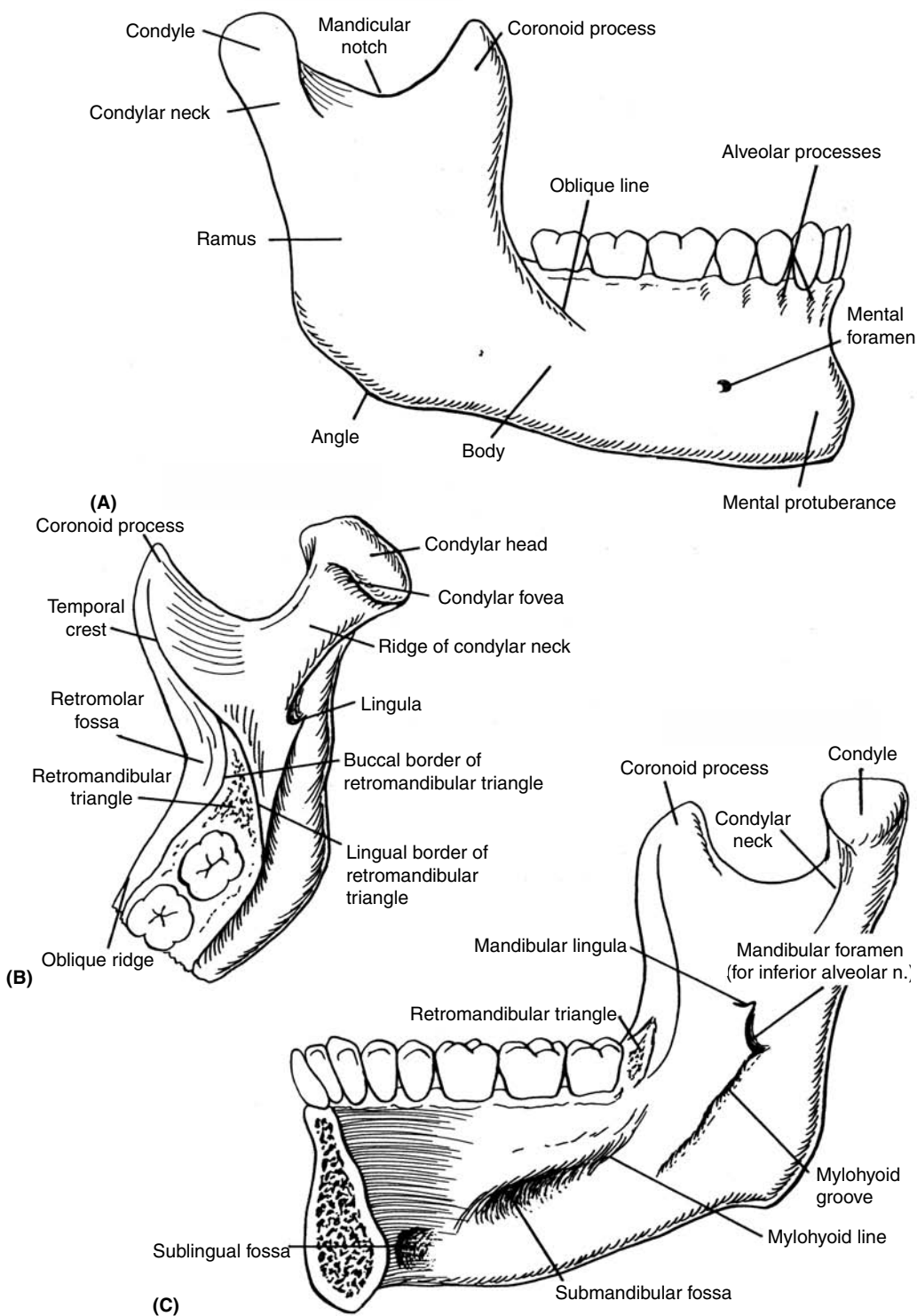


FIGURE 35 The mandible; **(A)** lateral view, **(B)** superior view, and **(C)** medial view.

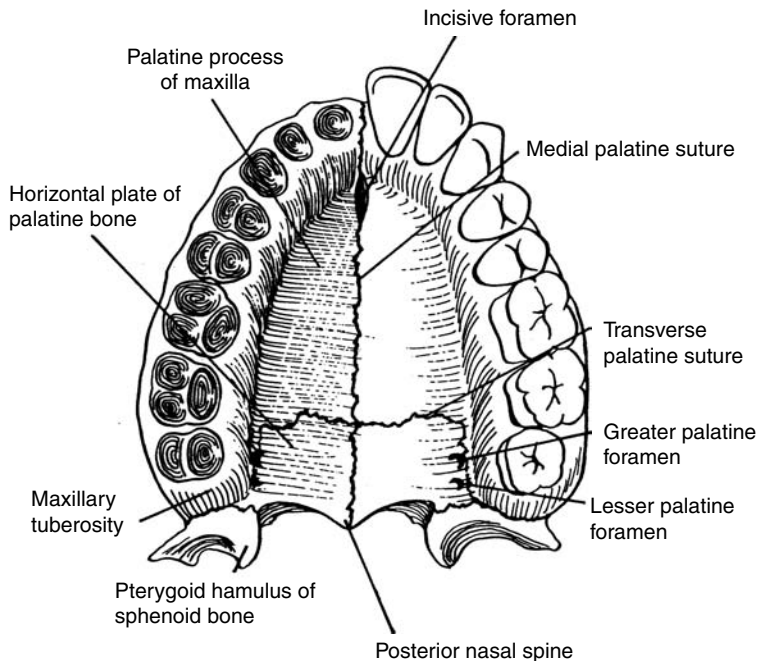


FIGURE 36 Inferior palatal view of the maxillary-palatine junction.

arteries, which pierce the medial wall of the orbit at approximately 24 and 34 mm from the rim. The anterior ethmoidal lies on the same plane as the cribriform plate and are a helpful guide to correct placing of a Le Fort III osteotomy. The posterior ethmoidal artery lying just anterior to the optic nerve may serve as a warning post that your medial wall dissection is entering a dangerous area.

The Cranial Facial Skeleton

The cranial facial skeleton is composed of 10 cranial and 15 facial bones. The cranial bones are one frontal and occipital, and two ethmoid, sphenoid, temporal, and parietal. The facial bones are all paired except for the vomer. The nasal, lacrimal, inferior nasal conchae, maxilla, zygoma, palatine, mandible and vomer comprise the bones of the face (Fig. 27).

The cranial bones are comprised of three layers an outer cortical table, and an inner cortical table separated by a vascular cancellous layer known as the diploic space. The thickness of the cranium averages between 6.8 and 7.6 cm and is thickest over the parietal region. The outer table is thicker than the inner table and when harvested at the layer of the diploic space provides excellent split thickness cranial bone graft for reconstructive purposes. When harvesting a split cranial graft, choose the location carefully. Grafts taken anterior to the hairline or future hairline may result in a visible depression. Additionally one should avoid harvesting over the sagittal midline to avoid potential laceration to the sagittal sinus. As the inner cortical table is relatively thin it is important not to lever with the osteotome against the inner table while raising the graft. This maneuver may produce a depressed skull fracture and underlying intracranial bleed (Fig. 28).

The separate bones of the cranium are fused along distinct suture lines, which remain open during the period of cranial growth. The frontal bone of the forehead joins the two parietal bones along the *coronal suture*. The central fusion point of these three bones is open at the time of birth and exists as the *anterior fontanelle*. The fontanelle averages about 2.1 cm in

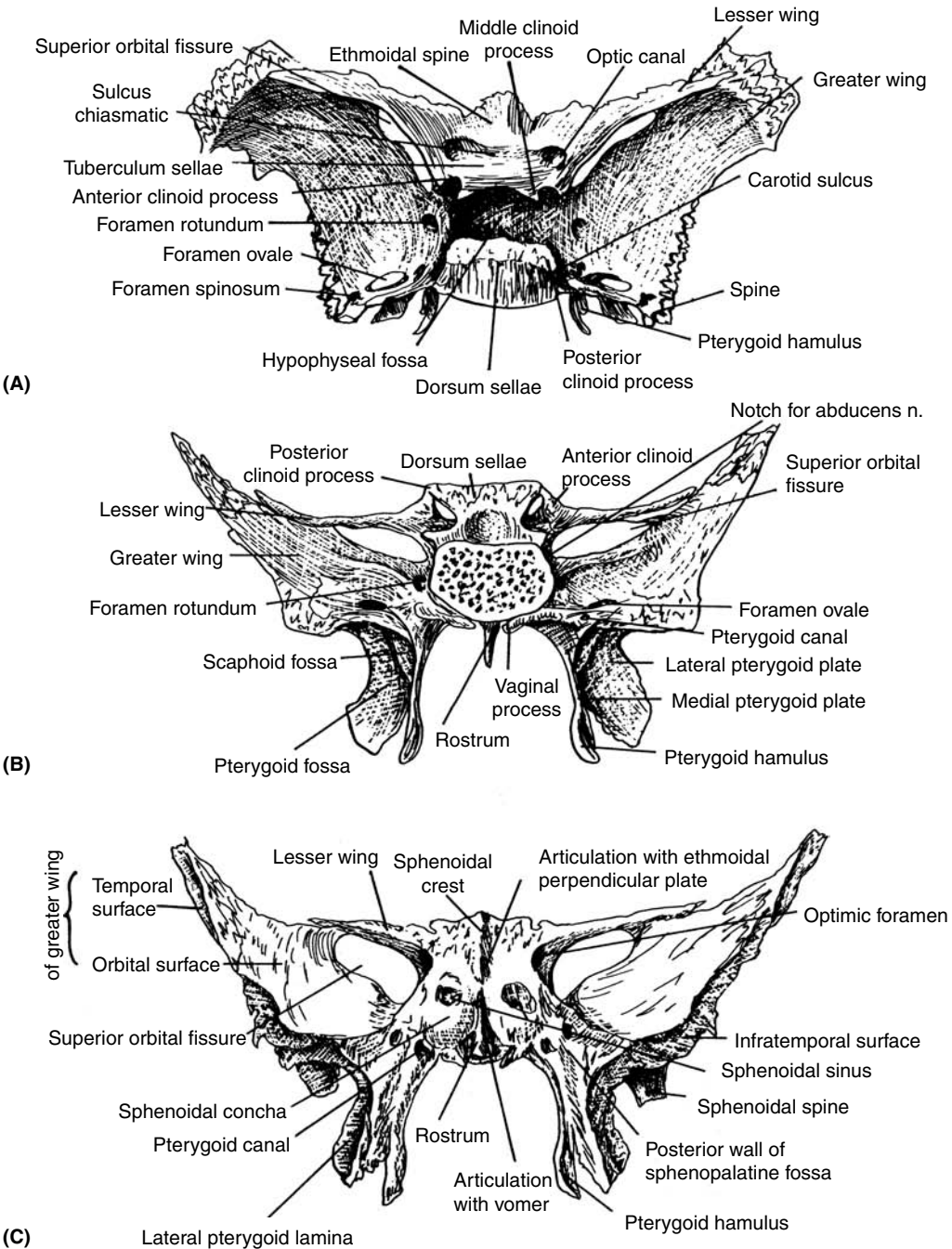


FIGURE 37 The sphenoid; **(A)** superior view, **(B)** posterior and slightly superior view, and **(C)** anterior and slightly inferior view.

size and closes at a mean of 14 months with a range of 4 to 26 months. On closure the fusion point of the three bones is known as the *bregma*. Although the frontal bone is a single bone at birth its intrauterine origin is from two bones, which fuse along the *metopic suture* line. The parietal bones fuse along the *sagittal suture* from the bregma to the *lambda*, the intersection between the sagittal suture and the *lambdoidal suture*, which joins the parietal bones to the

occipital bone. At birth the lambda is present as the *posterior fontanelle*, which measure from 0.5 to 1 cm and generally, closes by two months of age. The parietal bones fuse to the temporal bones at the lateral skull along the *squamosal suture* line (Fig. 29).

The internal cranial base is divided into three major segments, the anterior, middle, and posterior cranial fossa (Figs. 30 and 31). The *anterior cranial fossa* is composed of the frontal bone, the ethmoid and the lesser wing of the sphenoid. These bones are joined at the *sphenofrontal*, *sphenoethmoid*, and *frontoethmoidal* sutures. Early fusion of these sutures is responsible for some of the characteristic deformities seen in syndromic craniosynostosis such as Apert and Cruzon syndrome.

The *middle cranial fossa* is composed of the lesser wing of the sphenoid and the petrous portion of the temporal bone. These bones are joined at the *sphenosquamosal suture*. It is within the middle cranial fossa that the foramen, rotundum, ovale, lacerum, and spinosum transverse the sphenoid bone. The maxillary branch of V, the mandibular branch of V, the internal carotid artery, and the middle meningeal artery respectively run through this foramen.

The *posterior cranial fossa* is formed by the petrous portion of the temporal bones, and the occipital bone, which are joined by the lambdoid suture and its continuation the *occipitomastoid suture*. The foramen magnum and the jugular foramen are found in the posterior fossa. The internal jugular vein as well as cranial nerves IX, X, XI exit through the jugular foramen.

The facial bones are connected to the skull via the frontal sphenoid and temporal bones. Superiorly the frontal bone attaches to the facial bones along the *frontozygomatic*, *frontallacrimal*, *frontalmaxillary*, and *frontalnasal* suture from lateral to medial. The sphenoid bone attaches to the facial bones via the *sphenozygomatic suture* at the lateral wall of the orbit and the *sphenomaxillary suture* between the pterygoid process and the maxilla. The temporal bone attaches to the face at the posterior zygomatic arch at the *temporal zygomatic suture*. The Le Fort III osteotomy separates the cranial bones from the facial bones along these suture lines.

The individual cranial and facial bones are best appreciated by illustration rather than words. Examination of the following illustrations should provide the reader with an overall knowledge of the anatomy (Figs. 32–37).

Anatomic knowledge is the foundation of surgical practice. The craniofacial surgeon must first understand the normal then learn to perceive the abnormal by its anatomic context. Only then can the surgeon change what is, to what it should be.

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3 Anesthesia Considerations for Pediatric Craniofacial Surgery

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PREOPERATIVE EVALUATION

Infants and children who require craniofacial surgery present a special challenge to anesthesiologists. It is desirable to send many of these patients to the anesthesiologist for a preoperative visit prior to the day of surgery for evaluation particularly, if the anesthesiologists are unfamiliar with the patient. Many patients with craniofacial abnormalities (1) will require special techniques and equipment for securing an airway under anesthesia (Table 1). Preoperative evaluation will allow adequate time for a careful airway history and examination (2).

A history of obstructive sleep apnea or airway obstruction with supine positioning should be identified. Difficult mask ventilation and difficult intubation may be associated with high Mallampati class (Fig. 1) (3), short thyromental distance, mandibular hypoplasia, and micrognathia and/or limited neck mobility. Obesity, steroid use and a multitude of pediatric syndromes and diseases may also lead to increases in pharyngeal soft tissue and difficult ventilation or intubation (1). If the patient has had previous surgery, a preoperative visit may also allow time to obtain old anesthesia records, which may be a guide for airway management in the operating room. It has also been shown that prediction of intubation by direct laryngoscopy could be estimated by the radiographic classification of the mandibular deformity in patients with unilateral hemifacial microsomia (4).

Pediatric patients may have associated or sporadic coexisting diseases that will require optimization prior to surgery. Children with cardiac disease require special attention to recent echocardiographic findings, perioperative management of cardiac medications and subacute bacterial endocarditis prophylaxis. Patients with metabolic deficiencies may need special fluid management and tailoring of the American Society of Anesthesiologists (ASA) fasting guidelines (Table 2) (5) in the perioperative period. No matter what the associated syndrome, proper preparation with the anesthesiologist will aid in the intraoperative medical management of these patients. Previous airway surgery should also be an important consideration before the administration of anesthesia. Nasal, pharyngeal, and laryngeal tissue anatomy may be changed or hardware may be in place that may make airway management difficult.

Preoperative laboratory testing should be guided by associated conditions and the likelihood of perioperative blood transfusion. If there is a high probability of the need for transfusion with surgery, consideration should be given to drawing a preoperative sample for the blood bank and hemoglobin level. Infants between three and six months of age are well known to have a physiologic anemia secondary to the conversion of fetal hemoglobin to adult hemoglobin. Multiple studies have examined preoperative administration of erythropoietin to decrease the incidence of transfusion for repair of craniosynostosis (6–8). Normovolemic hemodilution may also be used as an adjunct to decrease the amount of red blood cells lost (9).

PREPARATION OF THE OPERATING ROOM

Beyond the standard equipment and monitoring required by the ASA, special preparation may be required for pediatric patients especially those with craniofacial abnormalities. The first step requires the selection of airway devices of multiple sizes. Sizing of masks, laryngoscope blades,

TABLE 1 Pediatric Syndromes Commonly Associated with a Difficult Airway

Apert syndrome
Beckwith–Wiederman syndrome
Cleft palate
Crouzon syndrome
Down syndrome (trisomy 21)
Fibrodysplasia ossificans progressiva
Freeman–Sheldon syndrome
Goldernhar syndrome (hemifacial microsomia)
Klippel–Feil syndrome
Mucopolysaccharidoses
Pierre Robin complex
Pfeiffer syndrome
Treacher–Collins syndrome

endotracheal tubes (cuffed or uncuffed), oral and nasal airways, and other airway adjuncts may be estimated by patient age and weight, but final selection needs to be individualized to each patient. The equipment setup should include at least one size above and one below the estimated size. This will help insure that potentially lifesaving airway equipment is readily available. It may also be advisable to have a laryngeal mask airway (LMA) of various sizes also available as use of the LMA has been added to the ASA emergency airway algorithm (Fig. 2). Preparation of a pediatric tracheostomy set should be considered for patients with a difficult airway.

After the preoperative airway examination, the anesthesiologist may decide that a flexible fiber optic bronchoscope will be used for tracheal intubation. Fiber optic bronchoscopes for tracheal intubation come in multiple sizes (Pentax Medical Company, Montvale, New Jersey, U.S.A.; Olympus America, Inc., Center Valley, Pennsylvania, U.S.A.; Karl Storz Endoscopy–America, Culver City, California, U.S.A.). The smallest sizes at 2.2 to 2.5 mm outer diameter do not have a side channel for the insufflation and are extremely flexible. The fiber optic bronchoscopes that are at least 2.7 mm in outer diameter are available with a side channel. The flexible fiber optic bronchoscope may be used through the oral or nasal route or through an LMA (10). Other available intubation equipment for known difficult airways includes, but is not limited to, the Bullard laryngoscope (11) (Circon-ACMI, Stamford, Connecticut, U.S.A.), video-optical intubation stylet (12) (Volpi Manufacturing, Auburn, New York, U.S.A.), and the Shikani Optical Stylet (Clarus Medical, Minneapolis, Minnesota, U.S.A.) (13).

INDUCTION OF ANESTHESIA AND AIRWAY MANAGEMENT

The choice of the induction of anesthesia will be determined by the preoperative airway assessment and the age of the patient. In older and compliant children, intravenous (IV) access is often established prior to induction of anesthesia. Establishing an IV in children is often preceded by the use of an oral, nasal, rectal, or intramuscular premedication. Topical anesthetics may also be helpful in placing an IV in infants and children. The advantages of

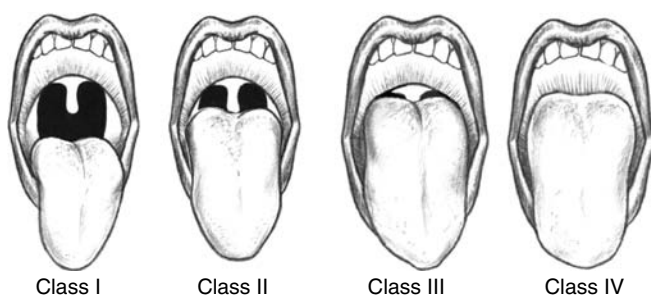


FIGURE 1 Mallampati class. *Source:* Illustrations provided by Linda Blue.

TABLE 2 American Society of Anesthesiologists Fasting Guidelines

Ingested material	Minimum fasting period (hours)
Clear liquids	2
Breast milk	4
Infant formula	6
Nonhuman milk	6
Light meal	6
Fried/fatty foods	8

IV access prior to induction include the ability to administer fluids, anesthetics, antisialogogues, and emergency medications.

If IV access is placed, there are multiple choices for the induction of anesthesia. Standard induction for presumed normal airways is usually performed with propofol, thiopental, etomidate, or ketamine for hypnosis plus a nondepolarizing muscle relaxant for intubation after mask ventilation has been established. Ketamine is often used for IV induction of anesthesia in patients with presumed difficult airways as it is known to have less respiratory depression, when compared with the other anesthetic agents (14). Dexmedetomidine is a relatively new hypnotic agent that may prove to be useful for fiber optic intubation in pediatric patients (15), but requires further evaluation. A combination of an opioid and a benzodiazepine may also be used with the advantage that reversal agents exist for both of these medications.

If the decision is made to induce anesthesia without IV access, then mask or inhalation induction must be employed. Sometimes, anesthesiologists will choose an inhalation induction even if IV access has been established. The main advantage of inhalation induction is that it is painless for the patient, but children and infants may still exhibit anxiety or combativeness with a mask induction. The three agents used currently for inhalation induction include nitrous oxide, sevoflurane, and halothane. Nitrous oxide is odorless and well tolerated, but lacks the potency for the complete induction of general anesthesia as a sole agent. It may be used in combination of sevoflurane or halothane, but is used in doses of 50% to 70% of the inhaled gases, which will significantly decrease the FiO_2 during induction. Sevoflurane has a faster onset than halothane and may have less depressant effects on the myocardium (16). The other advantage of inhalation induction is the easy titration of anesthetic agent with maintenance of patient respiration. The main disadvantage is the risk of laryngospasm or airway obstruction during induction of anesthesia. Intramuscular medications including atropine and succinylcholine may be given if laryngospasm occurs and may be predrawn in syringes prior to induction.

If mask ventilation is difficult, there are several airway maneuvers that may be helpful. Aside from the proper selection of equipment, proper patient positioning is important. Elevation of the head may not be necessary in infants secondary to the large occiput in these patients. A shoulder roll may also help in aligning the airway for good mask ventilation. At our institution, a tongue stitch may be placed with local and/or IV anesthesia to assist in extruding the tongue in patients with severe micrognathia requiring mask ventilation.

Intubation of the trachea may be performed by direct laryngoscopy or fiber optic laryngoscopy. Either procedure may be performed under sedation and/or topicalization with local anesthetic with spontaneous ventilation or after the induction of general anesthesia with spontaneous or controlled ventilation. A helpful maneuver utilized at our institution involves the use of a nasopharyngeal tube placed for oxygen and anesthetic agent while the fiber optic laryngoscope is used (17). This has allowed for a longer time for intubation during difficult cases.

The decision to plan the timing and technique for intubation should be determined prior to the induction of anesthesia with consideration of patient age, airway exam, and comorbidities. The risks and benefits of nasal versus oral intubation must also be considered, but this may also be predetermined by the operative site. Elective tracheostomy (18) may also be performed under mask anesthesia or after tracheal intubation. No matter which technique is chosen, properly securing the endotracheal or nasotracheal tube is of the utmost importance and may require suturing of the tube to the patient. Confirmation of tracheal tube placement

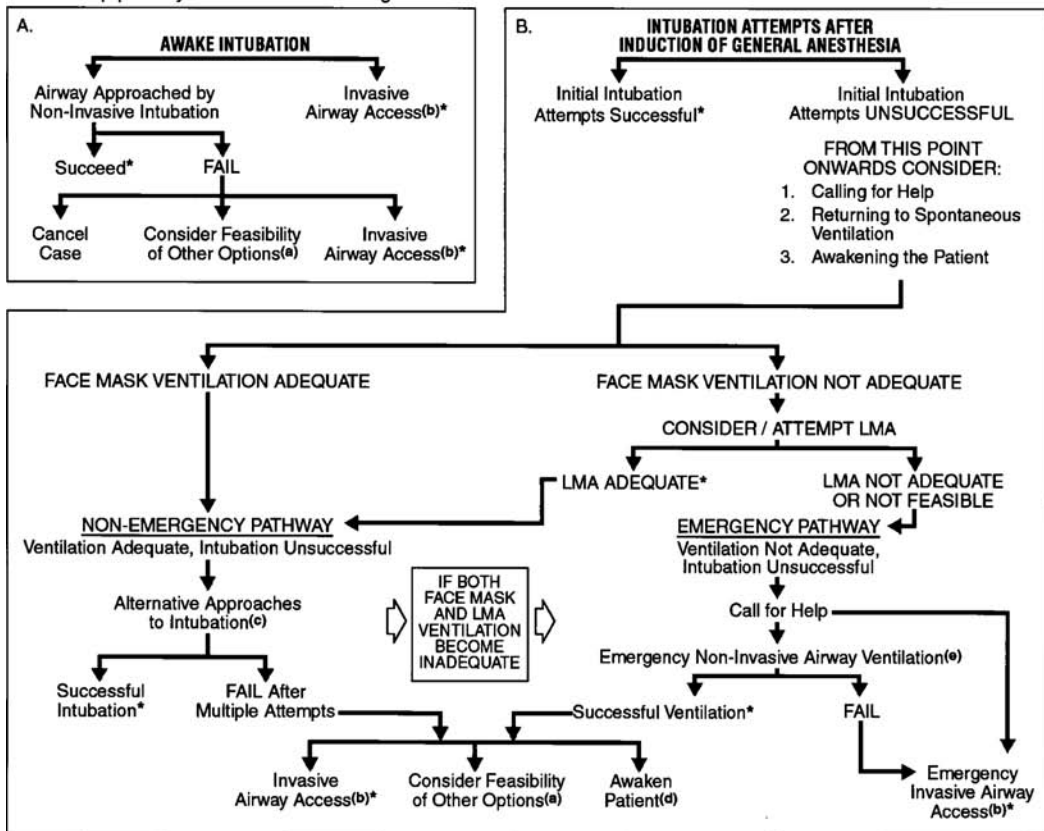


DIFFICULT AIRWAY ALGORITHM

1. Assess the likelihood and clinical impact of basic management problems:
 - A. Difficult Ventilation
 - B. Difficult Intubation
 - C. Difficulty with Patient Cooperation or Consent
 - D. Difficult Tracheostomy
2. Actively pursue opportunities to deliver supplemental oxygen throughout the process of difficult airway management
3. Consider the relative merits and feasibility of basic management choices:

- A. Awake Intubation vs. Intubation Attempts After Induction of General Anesthesia
- B. Non-Invasive Technique for Initial Approach to Intubation vs. Invasive Technique for Initial Approach to Intubation
- C. Preservation of Spontaneous Ventilation vs. Ablation of Spontaneous Ventilation

4. Develop primary and alternative strategies:



* Confirm ventilation, tracheal intubation, or LMA placement with exhaled CO₂

a. Other options include (but are not limited to): surgery utilizing face mask or LMA anesthesia, local anesthesia infiltration or regional nerve blockade. Pursuit of these options usually implies that mask ventilation will not be problematic. Therefore, these options may be of limited value if this step in the algorithm has been reached via the Emergency Pathway.

b. Invasive airway access includes surgical or percutaneous tracheostomy or cricothyrotomy.

c. Alternative non-invasive approaches to difficult intubation include (but are not limited to): use of different laryngoscope blades, LMA as an intubation conduit (with or without fiber optic guidance), fiber optic intubation, intubating stylet or tube changer, light wand, retrograde intubation, and blind oral or nasal intubation.

d. Consider re-preparation of the patient for awake intubation or canceling surgery.

e. Options for emergency non-invasive airway ventilation include (but are not limited to): rigid bronchoscope, esophageal-tracheal combitube ventilation, or transtracheal jet ventilation.

FIGURE 2 American Society of Anesthesiologists (ASA) emergency airway algorithm. Source: From Ref. 23.

can be established by auscultation of equal breath sounds, presence end-tidal carbon dioxide, fiber optic bronchoscopy, and chest X ray.

INTRAOPERATIVE MANAGEMENT

After the airway is secured, additional IV access and monitors are placed by the operating room team. If significant blood loss is expected, adequate IV or central access for the administration of fluids and blood products should be established. Central access may also be used for blood sampling. Use of an arterial line may also be considered for procedures where large quantities of blood loss are expected and for intracranial procedures. Proper IV access and arterial blood pressure monitoring are especially important for small infants and children undergoing procedures with large relative blood loss. Vigilant assessment of blood loss and circulating blood volume will be a helpful guide in fluid administration and management (19). Erythropoietin combined with normovolemic hemodilution has been successfully used to minimize the use of blood products during surgery for craniosynostosis (19). Use of a transcutaneous Doppler as an additional monitor may be considered for procedures with a high risk of venous air embolism. Solid and frequent communication between the anesthesia and surgical team will help improve the probability of a good outcome. This communication is particularly crucial during an acute situation of hemodynamic instability. At times, the surgical team may need to place warm soaked lap sponges over the surgical site, stop operating, and allow the anesthesiologist to volume resuscitate the patient.

POSTOPERATIVE MANAGEMENT

The anesthesiologist and surgeon should agree upon a plan for extubation. For most procedures, extubation will follow immediately after the conclusion of surgery. Extra care must be taken in patients who have undergone craniofacial surgery to ensure a successful extubation. Reintubation will often be more difficult in these patients as facial surgery will often distort the airway from anatomical changes, inflammation, edema, and bleeding. In addition, reintubation will risk disrupting the surgical repair with palate and pharyngeal procedures.

In order to decrease the risk of the need for assisted ventilation, drugs with respiratory depression should be carefully titrated, recovery from muscle relaxants should be carefully assessed, and respiratory function should be adequate. Infants and small children are at increased risk for postextubation croup secondary to their small airways especially at the narrowest point, the cricoid ring. The same equipment and personnel required for intubation is needed for extubation (2). Typically, extubation in the operating room after the procedure is safest. Skilled personnel (pediatric anesthesiologist and surgeon) and specialized equipment are immediately available. If there is concern about swelling around the endotracheal tube after a prolonged procedure, then a perioperative dose of steroids may be used. Facial edema does not always correlate directly with concomitant airway edema but in some circumstances may be an indicator. The use of local anesthetic for analgesia may decrease the need for opioid medications in the postoperative period. Additionally, use of an airway exchange catheter may be considered as an adjunct for extubation in patients who were difficult to intubate or who may be at risk for reintubation (20).

In some cases, the anesthesia and surgical teams may elect to keep the patient intubated to be extubated after further recovery from anesthesia and airway edema. Additionally, patients may remain electively intubated to allow for continued daily mandibular distraction by turning arms such as for micrognathia (21). In these circumstances, the patient will be transported to the pediatric intensive care unit (PICU) directly from the operating room or post-anesthesia care unit (PACU). Careful attention of the airway should be maintained during transport. At our institution, we maintain constant end-tidal CO₂ monitoring during transport and in all intubated PICU patients. A chest radiograph is obtained upon admission to the PICU to confirm correct tracheal tube placement. The same criteria for extubation are used as in the operating room as well as equipment and personnel available.

A prospective study on clinical outcome of perioperative airway and ventilatory management in children who underwent craniofacial surgery was conducted from 1999 to 2002 in 95 patients who underwent 99 procedures (22). Tracheal extubation was successful in the PACU in 58% of the patients and 42% received mechanical ventilation for up to 60 hours postoperatively in the PICU. Three percent of the patients required reintubation after failed extubation in the PICU. Patients who required fiber optic bronchoscopy for the initial intubation had a significantly greater length of intubation and mechanical ventilation when compared with patients who underwent direct laryngoscopy.

SUMMARY

Adequate preoperative evaluation is imperative for the anesthetic management of infants and children for craniofacial surgery. This will allow assessment of the airway and the potential for blood loss and proper preparation of the patient and operating room. This evaluation will guide the anesthesiologist as to the type of induction and the need for intraoperative monitoring and access. It is optimal if the same specialized personnel and equipment are readily available for the entire perioperative period which extends through recovery from anesthesia and extubation. Good communication between the entire perioperative team is essential in the care of these patients.

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4 Neurosurgical Considerations in Craniofacial Surgery

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INTRODUCTION

As the name of our specialty implies, the craniofacial surgeon will often be involved in cases where transcranial exposures, manipulation of the cranial vault, and the use of intracranial techniques are called for. For this reason, we must be more than just peripherally knowledgeable in the practical application of neurosurgical principles. In particular, we must be skilled in the identification and management of perioperative neurosurgical issues requiring urgent or emergent attention. No craniofacial team is complete without the active participation of a pediatric neurosurgeon during the preoperative assessment, the surgical intervention, and the postoperative follow-up. Below we will discuss some of the neurosurgical considerations that must be taken into account by both plastic surgeons and neurosurgeons in the management of the craniofacial surgery patient.

PREOPERATIVE CONSIDERATIONS AND PATIENT ASSESSMENT

Patients requiring intracranial procedures should be thoroughly evaluated by both plastic surgery and neurosurgery teams preoperatively. Detailed histories should be obtained and physical examinations performed. Whether these evaluations are performed simultaneously in the craniofacial clinic or independently sometime before surgery is immaterial—what ultimately matters is that a surgical plan incorporating the needs and goals of each team is developed. This requires direct communication between the plastic surgeons and neurosurgeons prior to surgery. In high-volume centers where the caseload of intracranial cases is significant and the same craniofacial and neurological surgeons work together regularly, preoperative discussion may be limited simply to elaborating upon any deviations from the normal established “routine.” Where there are rotating surgeons who staff the craniofacial clinic, however, or in centers where complex craniofacial cases are less common, the two teams must educate one another as to their respective practices. This includes mutual education regarding intraoperative positioning and equipment needs, desired incisions, and other relevant preferences. Such collaboration should occur far enough in advance of the surgical date to allow adequate time for preparation.

Attention to neurosurgical issues must begin during the preoperative assessment; specific preoperative workup must be tailored to each individual case. Any suspicion of intracranial communication of extracranial lesions must prompt an aggressive and thorough evaluation. In certain instances, the indication for performing the surgery itself is dependent on the information derived from imaging studies. Specific examples are discussed below.

Vascular Anomalies

Any suspicion of abnormal vascular anatomy that may be associated with a craniofacial anomaly must be completely investigated with computed tomography (CT) or magnetic resonance angiography prior to surgery.

Patients with *velocardiofacial syndrome* who have associated hypernasality following cleft palate repair and who are candidates for sphincter pharyngoplasty or pharyngeal flap surgery are at risk for vascular injury due to the aberrant anatomy of the internal carotid vessels associated with this disorder (Fig. 1) (1). These patients must undergo thorough intraoral examination, including inspection and palpation of the pharyngeal walls, and in some cases adjunctive magnetic resonance angiography of the head and neck, to document the anatomy of the vessels and alter the surgical plan accordingly (2,3).

Moyamoya, a progressive cerebrovascular occlusive disease, has been associated with the development of basal encephalocele (4,5). Failure to identify the vascular anomaly prior to an attempt at surgical correction of the cleft can have severe deleterious consequences.

Midline Masses

Any patient who presents with a congenital midline facial mass must be evaluated for the presence of intracranial extension. Dermoid cysts, gliomas, and frontonasal encephaloceles all represent lesions of varying degrees of differentiation that result from failure of regression of embryonic dural evaginations that extend through the foramen cecum to the prenasal space (6). If the tract persists and the foramen fails to close, any one of these lesions may develop. Up to 45% of midline dermoids noted at birth have been shown to have intracranial components (7), and up to 25% of congenital midline gliomas possess intracranial stalks (8). Whether the midline mass is a small lesion (e.g., a simple sinus tract) or a large frontonasal encephalocele consisting of herniated meninges and brain, detailed radiographic imaging is necessary to help plan a combined intracranial/extracranial approach with the neurosurgical team (Fig. 2).

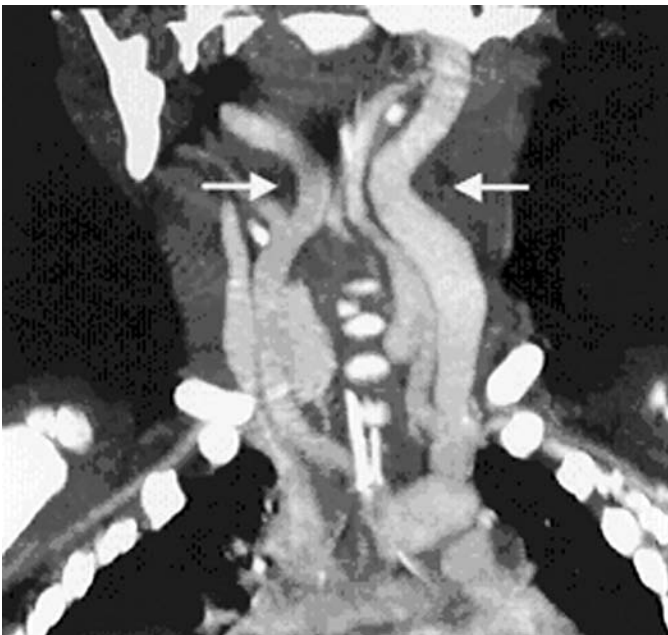


FIGURE 1 Coronal view reconstruction of a computed tomography angiogram in a patient with velocardiofacial syndrome reveals medial displacement of the internal carotid arteries at the level of the pharynx.



FIGURE 2 A large midline frontonasal encephalocele herniating through a Tessier #0/14 cleft presenting with hypertelorbitism, cleft nose, and cleft palate.

Craniosynostosis

There are varying reports on the incidence of elevated intracranial pressure (ICP) associated with craniosynostosis. Elevated ICP and developmental delay are seen more commonly in multiple-suture and syndromic craniosynostoses (9–11). When the deformity is mild and the indication for surgical intervention is not absolute, preoperative workup—including fundoscopic examination by a neuro-ophthalmologist to identify papilledema, CT scan to determine the presence of scalloping or thumbprinting of the inner table of the calvarium (Fig. 3A,B), and detailed neurological assessment to identify any neurological delay—may help stratify patient risk and determine the need for surgical intervention (12,13). Placement of an ICP monitor may be useful in determining the effects of synostosis on the ICP and to serve as a baseline for comparison to postoperative ICP levels (11,14).

INTRAOPERATIVE CONSIDERATIONS AND SURGICAL TECHNIQUES

The role of each surgical team should be well defined prior to the start of the procedure. As discussed above, patient positioning and preferred incisions should be predetermined and all necessary instruments and monitors should be immediately available. Additionally, the ancillary support team, including scrub technicians and circulating nurses, should be familiar with the instrumentation needs of both teams, so as to prevent intraoperative delays when any equipment is requested. Perioperative anesthetic care must be tailored to the individual patient and case, but intracranial surgery usually mandates continuous electrocardiogram monitoring, multiple venous lines, an intra-arterial catheter, and real-time capnography. In addition, continuous precordial ultrasound may be used to monitor ventricular wall motion and to detect the presence of air embolism (15).

Typically, the plastic surgeon will perform the initial dissection of the soft tissues of the scalp and the neurosurgeon will perform the craniotomy once the calvarium is exposed. The use of serial preoperative injections of erythropoietin has been shown to minimize the need for

perioperative transfusion (16). We continue to favor subperiosteal dissection and bone wax to obtain hemostasis from emissary vein bleeding, but initial suprapariosteal dissection may limit blood loss during the exposure. This technique, however, may have ramifications for the vascularity of bone flaps after they are replaced.

Once the calvarium is exposed, the neurosurgeon and plastic surgeon decide upon the optimal placement of burr holes and the design of the craniotomy and/or craniectomy. Burr holes should be paramedian to prevent injury to the midline sagittal sinus and should be placed above the cephalic extent of the frontal sinus. Extradural dissection is performed through the burr holes to free the dura from the overlying vault. Great care must be taken when this dissection is performed over the sagittal sinus. The dura tends to be most adherent to the skull at the cranial sutures; the additional force needed for dissection in these areas must be balanced against the risk for dural tears.

The bone flaps are cut using a drill with a guarded footplate. Once the bone is removed, the dura is irrigated and hemostasis is obtained using bipolar electrocautery. An immediate assessment should be made regarding ICP. If the brain is too "tight," the anesthesiologist must institute corrective measures. These include head elevation, hyperventilation, and administration of steroids and an osmotic diuretic, such as mannitol. Any dural tears that are identified are immediately repaired with nonabsorbable suture. At closure, dural repairs should be reinforced to add an additional layer of protection against cerebrospinal fluid (CSF) leak. We prefer the use of pericranial flaps or grafts covered with fibrin tissue glue (17). The dura must be intermittently irrigated throughout the case so as to prevent desiccation from prolonged exposure. Of course, in all intracranial cases the brain must be handled with extreme care. Excessive pressure during retraction or indelicate movements may rupture fragile veins and cause subdural hemorrhage.

The exposures required for frontoorbital advancement (FOA), exploration of the orbit, and temporoparietal barrel staving all require aggressive brain retraction. For FOA, the anterior floor of the middle cranial fossa must be fully exposed in order to protect the intracranial contents as the infratemporal fossa osteotomy is made and carried medially (Fig. 4). In order to achieve this, the pterion must be reduced, usually with a rongeur, and the dura freed from the cranium. Great care must be taken here, as the dura may be tightly adherent to the greater wing of the sphenoid and dural tears in this area are difficult to repair. Any surgical manipulation in the area of the posterior orbit must not place any traction on the optic nerve as it exits the optic foramen of the sphenoid. Likewise, traction injury of the olfactory bulbs or fila olfactoria can result in permanent deficits. When lateral dissection is performed for temporoparietal barrel staving, the middle meningeal artery is at risk along the floor of the middle cranial fossa.

Elevation in ICP brought about by reduction of the anteroposterior, biparietal, or vertical dimensions of the cranial vault can have profound negative consequences. Acute increases in ICP can elicit a Cushing reflex, manifest as a sudden onset of bradycardia. This phenomenon is

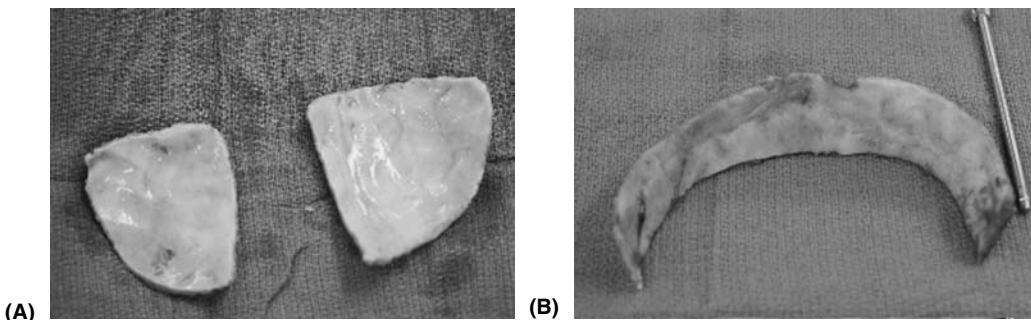


FIGURE 3 Right and left frontal bone flaps (A) and calvarial segment including excised coronal suture (B) in a patient with bilateral coronal and sagittal synostosis. The beaten copper appearance of the inner table of the skull indicates a history of elevated intracranial pressure.



FIGURE 4 A wide malleable retractor shields the frontal and temporal lobes, while the reciprocating saw makes an osteotomy through the sphenoid bone. The retractor is placed on the floor of the middle cranial fossa to completely protect the temporal lobe. A smaller ribbon retracts the orbital contents.

seen in the correction of scaphocephaly as well as during remodeling for reduction of turricephaly. As autoregulatory mechanisms correct the elevated ICP, the bradycardia may resolve, allowing additional correction. If the bradycardia persists, the correction must be released and recalculated. The anesthesiologist must be informed when these maneuvers are being performed and anticipated appropriate treatment measures.

In cases where the intracranial volume is increased (e.g., FOA for bicoronal synostosis), a void space may be created between the dura and the newly positioned inner table of the mobilized calvarial segment (Fig. 5). The brain should expand to occupy this space with time. In the immediate perioperative period, however, it poses an infectious risk. We therefore advocate the use of tacking sutures to affix the dura to the advanced bony segment to fill the void with soft tissue.

When the intracranial portions of these major craniofacial cases are completed, the bone flaps are replaced after remodeling and repositioning. We favor the use of absorbable plates and screws to secure the flaps in place in the pediatric population (Fig. 6) (18,19). This particular hardware is pliable when irrigated with saline solution warmed to 80°C. When using this irrigation to shape plates that are in situ, one must take great care not to scald the dura.

Closure of the scalp is usually completed by the plastic surgery team. It falls upon the plastic surgeon, therefore, to perform a final intraoperative examination to ensure complete hemostasis and absence of any CSF leakage. The scalp flaps are secured over closed suction drains, and the wounds are reapproximated in layers. Simple head dressings are kept in place overnight.

INTRAOPERATIVE EMERGENCIES

Hemorrhage

There are numerous potential sources of major bleeding in complex craniofacial cases. These include the scalp, the skull (via emissary veins as well as from corticotomies), arterial vessels (e.g., middle meningeal artery), and the venous sinuses. Because of this, and because



FIGURE 5 Barrel staving increases the intracranial volume in this patient with metopic suture synostosis characterized by decreased bitemporal diameter.

infants and children can rapidly lose a significant percentage of their total circulating blood volume, cross-matched blood products must be immediately available prior to the start of the case. As discussed above, we also support the hematocrit preoperatively through the use of serial erythropoietin injections, particularly during the period of physiologic infant anemia (20).

Bleeding from the scalp is controlled by opening the incision segmentally and by placing clips on the incised edges. Electrocautery should be avoided to prevent alopecia. As scalp flaps are raised, bleeding from the passage of emissary veins through the scalp is treated with bone wax. Again, suprapariosteal dissection may limit bleeding during this part of the case. All craniotomy sites will bleed, but application of bone wax should be sufficient to provide hemostasis. It should be noted, however, that bone wax has been shown to impede bone growth (21); it should therefore be judiciously used.

The most severe and potentially lethal hemorrhage comes from sinus injury. The sagittal sinus is the most commonly exposed and involved reservoir due to its midline position. It can be lacerated during placement of burr holes, during the craniotomy, or while attempting to remove the cut bone plates. When performing strip craniectomy for scaphocephaly or pansynostosis involving the sagittal suture, the sinus is at even greater risk. Adhesions of the dura to the calvarial vault, especially in the region of the lambdoid suture, can be particularly problematic. Any bleeding from the sinus must be treated immediately and aggressively. Failure to recognize the sinus injury or quickly obtain hemostasis can result in death, as the sinus may drain up to 50% of total cerebral arterial inflow (22). If the sinus is not fully exposed when the bleeding begins, cotton gauze can be placed in the area of injury until the craniotomy is extended. The head of the bed should also be elevated to decrease ICP. Once the source of bleeding is completely exposed, gentle pressure is applied and the bipolar electrocautery is used to obtain hemostasis. Alternatively, a flap of dura can be turned over to cover a tear overlying the sinus. The sinus is kept moist after hemostasis is obtained to prevent desiccation. Unless absolutely necessary, the sinus should not be packed as a means of controlling the hemorrhage. Sinus thrombosis, which



FIGURE 6 A left frontal bone flap is fixed in place using absorbable plates and screws.

has been associated with subsequent cerebral edema and intracerebral hemorrhage, is the likely outcome of this measure (23).

Venous Air Embolism

Air may be introduced into the venous drainage system anytime the surgical site is above the level of the heart and collapsible veins are exposed to air due to a pressure gradient favoring air entry into the venous system over venous bleeding (24). The sitting position in pediatric neurosurgery cases has been shown to have an associated incidence of air embolism between 30% and 80% (25,26). A subset of these patients will become hemodynamically unstable (27). Awareness of this risk helps guide patient positioning during the procedure. The diagnosis of air embolism can be made following an acute drop in end-tidal partial pressure of expired carbon dioxide of at least 5 mmHg, with or without associated hypotension or arrhythmia may also be present. The most sensitive monitors that aid in diagnosis are transesophageal echocardiogram and the precordial Doppler probe (24,28). Once identified, air embolism must be immediately treated with elimination of the source of air entry and replacement of intravascular volume. Additional measures include positive end-expiratory pressure, administration of 100% oxygen, and bilateral digital pressure on the jugular veins. Continued postoperative mechanical ventilation may be indicated (29,30).

POSTOPERATIVE MANAGEMENT AND COMPLICATIONS

Patients must be monitored in the pediatric intensive care unit during the postoperative period until they are clinically stable. Hemodynamic parameters should be followed in real time, hourly neurological examinations should be performed, and baseline blood counts, electrolyte levels, and coagulation studies should be obtained and repeated as necessary. The surgeon must be vigilant for signs of developing complications, the more common of which are described below.

Infection

Perioperative parenteral antibiotics are administered to prevent infection. Infectious risk is the highest in patients where communication between the paranasal sinuses and intracranial cavity is established, as in monobloc advancement and facial bipartition (31). Knowledge of surgical landmarks is important to avoid entering the nasal cavity during procedures where mobilization of the midface is not required (e.g., FOA).

Hemorrhage

It is not uncommon for patients to lose greater than one-half of their blood volume during surgery. Because this is entirely replaced by transfusion, particular attention should be paid to repleting coagulation factors and monitoring for coagulopathy and bleeding complications. Hemorrhage may be heralded by increasing sanguinous output from closed suction drains. Any change in mental status or neurological exam might indicate an intracerebral hemorrhage and necessitates urgent radiographic evaluation. Evidence of intracranial bleed on postoperative CT scan mandates an emergent return to the operating room to decompress the intracranial space. Any abnormalities in coagulation parameters must be treated concomitantly.

Elevated ICP

Increased ICP may be due to cerebral edema, disruption of ventricular drainage, sinus thrombosis, or secondary to a decrease in intracranial volume. Treatment must be directed at the underlying cause. Any concern that ICP may be elevated in the postoperative period should prompt placement of an intracranial monitor at the end of the case. If ICP increases acutely in the postoperative period, an ICP monitor can be placed at the bedside.

Treatment of increased ICP includes head elevation, fluid restriction, controlled hyperventilation, and placement of a lumbar drain. Osmotic diuretics and hypertonic saline are useful pharmacological agents. Corticosteroids may also be beneficial. Patients with elevated ICP refractory to conventional treatment may be treated with high-dose barbiturates or may require operative intervention to decompress the brain (32–35).

CSF Leak

Leakage of CSF may appear in the immediate or delayed postoperative period. A leak may manifest as rhinorrhea or as increasing output from closed suction drains. Treatment options vary from aggressive to expectant, but most leaks will seal with time. A high output leak that is detected in the immediate postoperative period may indicate a significantly sized dural tear that was not recognized during the surgery or an inadequately repaired defect in the dura. In this situation, operative exploration is indicated to close and patch the leak. Healing of a leak can be hastened by placement of a lumbar drain (36).

SIADH

The syndrome of inappropriate release of antidiuretic hormone (SIADH) is characterized by hyponatremia, hypotonia (decreased serum osmolality), and increased urine osmolality and sodium concentration. SIADH is commonly observed after head injury, including injuries of an iatrogenic nature. Symptoms may include headache, nausea, vomiting, confusion, and convulsions or coma. The fundamental treatment of SIADH is based on the principle of fluid restriction to correct the hyponatremia and pharmacological prevention of urine concentration (37). There is recent evidence, however, that the hyponatremia seen in postoperative craniosynostosis patients is a manifestation of cerebral salt wasting syndrome (CSWS), not SIADH (38). The difference is significant in that, unlike SIADH, CSWS is managed by fluid resuscitation, often with normal saline. If postoperative metabolic disturbances are detected, the appropriate diagnosis must be made to render effective treatment.

SUMMARY

Comprehensive care can be administered to pediatric craniofacial patients with complex surgical problems in a safe manner. With appropriate cooperation and communication between the neurosurgical and plastic surgery services, excellent outcomes with minimal complications can be expected. The craniofacial team must be familiar with the neurosurgical considerations discussed above to provide the best care possible to the patients it serve.

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5

Restoring Facial Shape in Face Lifting: Anatomic Considerations and the Role of Skeletal Support in Facial Analysis and Midface Soft Tissue Repositioning

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INTRODUCTION

While skeletal support of the human face changes little from early adulthood through middle age, facial shape changes greatly. The morphology noted in the aging face results primarily from volumetric soft tissue deflation and facial fat descent in relation to the underlying skeletal pillars of the midface. The skeletal pillars that support the soft tissues of the midface are located superolaterally and include the zygomatic eminence, the lateral and infraorbital rim, the zygomatic arch, and the ramus of the mandible. In youth this skeletal support system forms a foundation, which fixates facial fat through the retaining ligament system that is situated overlying the parotid, anterior border of the masseter and malar eminence. As this ligamentous support becomes attenuated with age, fat descends, and in conjunction with deflation, facial shape changes. The youthful face tends to appear angular in contour, while in middle age facial fat becomes situated anteriorly and inferiorly in the face, and facial configuration becomes squarer, with little differentiation between the malar eminence and submalar fat. The underlying skeletal support not only influences the appearance of the face in youth but also affects facial shape in middle age, and is greatly affected by the loss of volumetric highlights overlying the midfacial skeletal pillars. Skeletal support therefore affects the surgeon's ability to restore facial shape, and is one of the key elements to consider in aesthetic treatment planning for facial rejuvenation.

The works of Skoog (1) and Mitz and Peyronie (2) enlightened plastic surgeons to the possibility of repositioning descended facial fat to the anatomic position of youth, providing an alternative to skin envelope tightening to enhance contour in the aging face. Repositioning descended facial fat reestablishes the volumetric highlights noted in youth thereby enhancing facial structure and support. In my opinion, restoring facial shape through facial fat repositioning is a better aesthetic concept than attempting to tighten a loose face. To consistently improve facial shape in face lifting requires the surgeon accurately understand the anatomic changes that have occurred in aging for a particular patient, and appreciate the importance of underlying skeletal support in formulating a treatment plan, incorporating one's aesthetic vision into a surgical destination which is appropriate for a specific patient.

ANATOMIC CONSIDERATIONS

Soft Tissue Anatomy

The anatomic arrangement of the facial soft tissues as a series of concentric layers is a key to the safety of rhytidectomy procedures. This concentric arrangement allows dissection within one anatomic plane to proceed completely independently from structures lying within another anatomic plane. The fascial layers consist of (i) skin; (ii) subcutaneous fat; (iii) superficial musculoaponeurotic system (SMAS) (superficial facial fascia); (iv) mimetic muscles;

(v) parotidomasseteric fascia (deep facial fascia); and (vi) the plane of the facial nerve, parotid duct, buccal fat pad, and facial artery and vein (3–7).

To master sub-SMAS dissection, the surgeon must understand the following anatomic components of facial soft tissue anatomy (8):

1. Structures within each layer are anatomically constant, despite variations in the thickness of the various layers from patient to patient.
2. On a two-dimensional basis, the facial nerve exhibits a variety of branching patterns, but on a three-dimensional basis, the facial nerve always lies within a specific anatomic plane. This anatomic arrangement allows the surgeon to perform extensive sub-SMAS dissection safely, as long as the dissection proceeds at a level superficial to the plane of the facial nerve.
3. There is significant variability in the thickness of the superficial fascial layer (SMAS). This variability of SMAS thickness is obvious from patient to patient. Also, the thickness of the SMAS will vary from one region of the face to another. Overlying the parotid gland, within the temporal region (temporoparietal fascia) and within the scalp (galea), the superficial fascia (SMAS) represents a substantial, discrete layer. As the superficial fascia is traced anteriorly in the face, overlying the masseter, buccal fat pad, and into the malar region, the SMAS tends to become thinner and less substantial. To elevate the superficial fascia in these areas requires precise dissection, so that the flap is thick enough to be useful in facial contouring.
4. The muscles of facial expression are arranged in four anatomic layers that overlap one another. The muscles that are encountered in face lifting; including the platysma, orbicularis oculi, zygomaticus major and minor, and risorius muscle are all superficially situated mimetic muscles. This is in contrast to deeply situated mimetic muscles such as the buccinator and mentalis muscle. Most of the muscles of facial expression lie superficial to the plane of the facial nerve. Because these muscles are superficial to the plane of the facial nerve, they receive their innervation along their deep surfaces. The only muscles within the facial soft tissue architecture, which lie deep to the plane of the facial nerve, are the mentalis, buccinator, and levator anguli oris muscles. Because these muscles lie deep to the plane of facial nerve, they receive their innervation along their superficial surfaces (9).
5. The muscles of facial expression which are situated superficially within the facial soft tissue architecture and are involved in the movement of facial skin are invested by the superficial fascia. By investiture, we mean the superficial fascia lines both the superficial and deep surfaces of these muscles. Because these muscles are invested by superficial fascia, this SMAS–mimetic muscle complex forms a single anatomic and functional unit which work together to move facial skin during animation.
6. Deep to the SMAS–mimetic muscle complex lies the deep facial fascia. The deep facial fascia represents a continuation of the superficial layer of the deep cervical fascia cephalad into the face. Where this fascial layer is identified, it is given specific nomenclature. Overlying the parotid gland, the deep fascia is termed “parotid fascia” or “parotid capsule”; overlying the masseter muscle, it is termed “masseteric fascia,” and in the temporal region, it has been termed “deep temporal fascia.” *The significance of the deep facial fascia is that all the facial nerve branches within the cheek lie deep to the deep facial fascia.* Typically, these nerve branches course deep to the deep fascia until they reach the muscles of facial expression which they innervate, at which point they penetrate the deep fascia to innervate these mimetic muscles along their deep surfaces (Fig. 1) (4).

In an overview of the architectural arrangement of the facial soft tissue, the essential point is that there is a superficial component of the facial soft tissue, which is defined by the superficial facial fascia and includes the SMAS and those anatomic components that move facial skin (including superficially situated mimetic muscle invested by SMAS, the subcutaneous fat, and skin). This is in contrast to the deeper component of the facial soft tissue, which is defined by the deep facial fascia and those structures related to the deep fascia (including the relatively fixed structures of the face, such as the parotid gland, masseter muscle, periosteum of the facial bones, and facial nerve branches). As the human face ages, many of

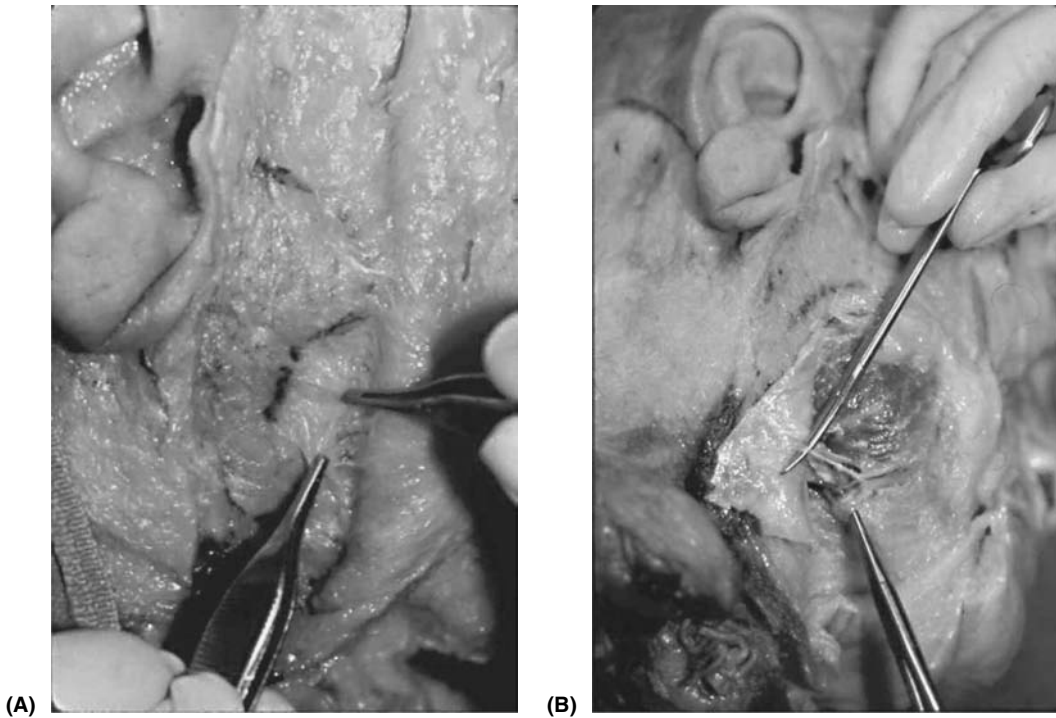


FIGURE 1 (A) Cadaver dissection following superficial musculoaponeurotic system-platysma elevation within the cheek, exposing the underlying parotid gland, anterior border of parotid (marked in ink), and parotidomasseteric fascia (held with forceps). The surgical significance of the parotidomasseteric fascia is that the facial nerve branches within the cheek are always deep to this anatomic layer. (B) Cadaver dissection following elevation of the parotidomasseteric fascia exposing the underlying masseter muscle and the marginal mandibular nerve as it crosses the facial artery and vein. *Source:* From Ref. 4.

the stigmata that are typically seen in aging relate to a change in the anatomic relationship which occurs between the superficial and deep facial fascia. With aging, facial fat descends in the plane between superficial and deep facial fascia, thereby justifying reelevation of fat through sub-SMAS dissection to improve facial shape.

Retaining Ligaments

The communication between the superficial and deep facial fascia occurs at the level of the retaining ligaments (4,10). These structures fixate facial soft tissue in normal anatomic position, resisting gravitational forces. Retaining ligaments exist within specific localities of the face, including (i) the malar area, where they are termed “zygomatic ligaments,” (ii) extending along the anterior border of the masseter, forming the masseteric cutaneous ligaments, (iii) overlying the parotid gland, forming the parotid cutaneous ligaments, and (iv) in the parasymphysial and symphyseal region of the mandible, where they are termed “mandibular ligaments” (Fig. 2).

There are two types of retaining ligaments, as defined by their origin. First, there are osteocutaneous ligaments, which are a series of fibrous bands that run from periosteum to dermis. The zygomatic and mandibular ligaments are examples of these structures.

A second system of supporting ligaments is formed by a coalescence that occurs between the superficial and deep facial fascia in certain regions of the face (parotid cutaneous ligaments, masseteric cutaneous ligaments). These fascial connections, which fixate both superficial and deep fascia to underlying fixed facial structures, such as the parotid gland and masseter muscle, similarly lend support against gravitational forces through fibrous septa that extend into the dermis.

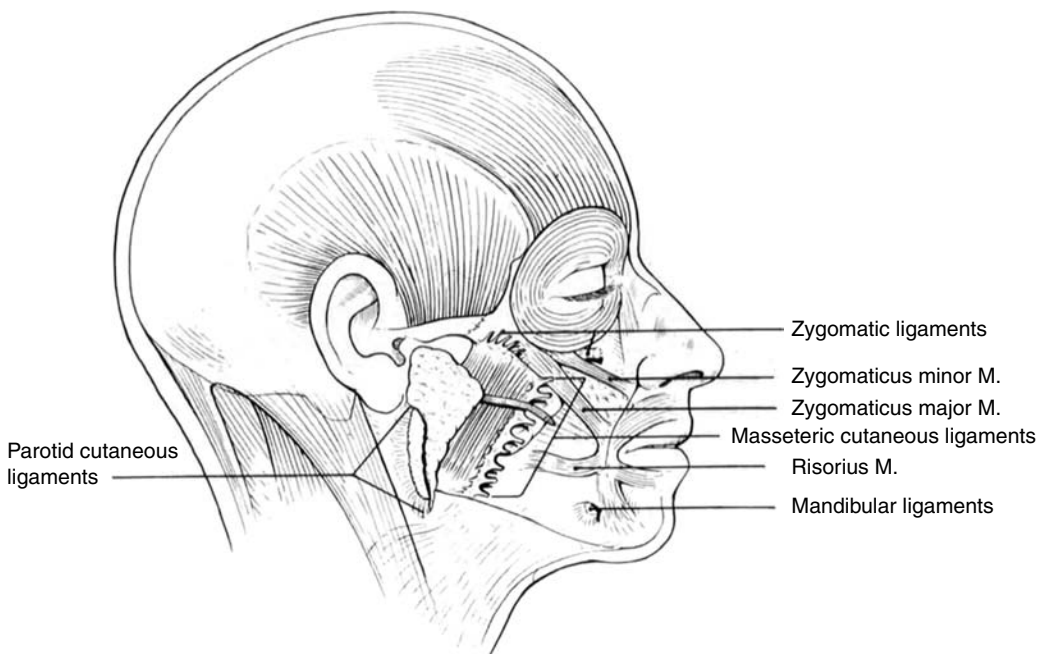


FIGURE 2 Facial soft tissue is supported in a normal anatomic location by a series of supporting ligaments. The zygomatic and mandibular ligaments are examples of osteocutaneous ligaments that originate from periosteum and insert directly into dermis. The masseteric cutaneous ligament and the parotid cutaneous ligaments are formed as a condensation between the superficial and deep facial fascias. Rather than originating from periosteum, these ligaments originate from relatively fixed facial structures such as the parotid gland and the anterior border of the masseter muscle. Attenuation of support from the retaining ligaments is responsible for many of the stigmata seen in the aging face. *Source:* From Ref. 4.

In the evolution of midface aging, the zygomatic and masseteric cutaneous ligaments bear particular attention. The zygomatic ligaments originate from the periosteum of the malar region. These ligaments exist as a series of fibrous septa that begin laterally in the region where the zygomatic arch joins the body of the zygoma and percolate throughout the malar pad (fibrous McGregor's patch). A particularly stout ligament is noted to originate along the most medial portion of the zygoma, medial to the zygomaticus minor. The zygomatic ligaments fixate the malar pad to the underlying zygomatic eminence in the youthful face.

Support of the soft tissues of the medial cheek is provided from a series of fibrous bands that extend along the entire anterior border of the masseter muscle. These fibers, termed "masseteric cutaneous ligaments," are identified superiorly in the malar area where they mingle with the zygomatic ligaments and extend along the anterior border of the masseter as far inferiorly as the mandibular border. These fibers represent a coalescence between the superficial and deep fascia, extending from the masseter muscle vertically to insert into the overlying dermis. These fibers support the soft tissues of the medial cheek superiorly above the mandibular border.

CHANGES IN FACIAL SHAPE WITH AGING

As the human face ages, facial shape changes. Morphologic facial changes are multifactorial. Some of these (changes) are straightforward to address, while others remain difficult technical challenges. A paradox for me has always been that facial anatomy (in terms of basic soft tissue architecture) is essentially unchanged from youth to middle age, but facial appearance changes greatly over time and is patient specific. Although each face ages differently, there are common themes noted in all aging faces.



FIGURE 3 (A) Age 25 years. The youthful face is full of well-supported facial fat, typically located overlying the malar eminence and along the lateral cheek, overlying parotid and masseter. This is associated with a concavity or depression overlying the buccal recess just anterior to the masseter. The combination of fullness in the malar region and lateral cheek associated with a concavity overlying the buccal recess accounts for the angular, tapered appearance of the youthful face. (B) Age 55 years. Thirty years later, the aesthetic effect of the descent of facial fat become obvious, resulting in a change in facial shape. Typically, faces in middle age are square in their configuration with little differential between malar highlight and midfacial fat. As facial fat is situated more inferiorly in the face, the face appears visually longer. The aesthetic consequences of a change in facial shape with aging are as important as the depth of the nasolabial fold and facial jowling. For me, an improvement in facial shape remains one of the primary goals in facial rejuvenation. *Source:* From Ref. 11.

Descent of Facial Fat

As the human face ages, facial fat descends and with it facial shape changes. Typically, the youthful face is full of well-supported fat. Volumetric highlights are located within facial aesthetic subunits that have a high density of retaining ligaments (malar, preparotid, orbital rim) and serve to fixate this volume of fat to underlying structures. Juxtaposed to the volumetric fullness (or convexity) of the malar and preparotid region is commonly a concavity within the submalar region, overlying the buccinator muscle and buccal recess. The combination of fullness in the malar region and lateral cheek, associated with submalar concavity and a well-defined mandibular border, accounts for the angular, tapered appearance of the youthful face.

With aging, facial fat descends and facial shape changes. In middle age, as ligamentous support becomes attenuated, facial fat volumetrically becomes situated anteriorly and inferiorly in the cheek, producing a facial contour that is squarer in configuration with little differential between malar highlights and midfacial fat. As facial fat is situated inferiorly in the face, older faces appear vertically longer than young faces (Fig. 3) (11).

Volume Loss and Facial Deflation

Young faces are full of well-supported facial fat. Over time, deflation of facial fat occurs, and tends to be most apparent in regions of the face with a high density of retaining ligaments



FIGURE 4 (A) Preoperative appearance of a 42-year-old patient with early facial aging resulting primarily from deflation. Note the hollowing effect along the infraorbital and lateral orbital rim and prepatrid region, which are regions of high ligamentous density. (B) Postoperative result following face and neck lift. Note, as anteriorly situated fat is brought into the upper lateral midface, it fills the areas of deflation, thereby blunting the lines of demarcation between aesthetic subunits that develop with age. Note also the change in facial shape, which now appears more structured and supported following facial fat repositioning. *Source:* From Ref. 14.

(12,13). For this reason, the areas that are noted to be volumetrically full in youth (malar, prepatrid, lateral and infraorbital rim, lateral chin) become volumetrically deflated in middle age. With facial deflation, soft tissue becomes less supported and therefore appears lax. Youthful faces have a smooth blending of contour between the aesthetic subunits of the face. Middle-aged faces, secondary to both deflation and facial fat descent, develop lines of demarcation between one region of the face and another which is intuitively identified as old. As part of the aesthetic treatment plan to improve facial shape, repositioning descended soft tissue into areas of facial deflation improves shape, not only by restoring volume to the position noted in youth but also serves to blunt the lines of demarcation which develop with aging (Fig. 4). Volumetric addition through autologous fat injection, dermal fat grafts, or other injectable soft tissue fillers are secondary agents that can be useful in augmenting areas of facial deflation (15,16).

Radial Expansion

Not all facial aging is vertical and a major challenge in facial rejuvenation is the radial expansion of facial soft tissue, which occurs along specific areas of the midface. In youth, the skin and underlying subcutaneous fat are densely attached to the deep facial fascia by reticular fibers that transverse between skin, subcutaneous fat, superficial fascia, and insert into the deep fascia and facial musculature. Over time, with prolonged animation such as smiling, the skin along the nasolabial line is forced deep to the subcutaneous fat, positioned lateral to the nasolabial fold, attenuating these reticular attachments (12,13). Prolonged animation therefore forces the skin and fat lateral to the nasolabial fold to expand radially and prolapse outward from the facial skeleton, accounting for much of the nasolabial fold prominence in the aging face. Radial expansion lateral to the oral commissure and marionette

line similarly accounts for some degree of jowl prominence in many middle-aged patients. Radial expansion is technically difficult to correct, as there are few surgical solutions to reestablish the retinacular attachments between skin, subcutaneous fat, and deep fascia. Because of this technical difficulty, incomplete correction in these regions is commonly noted following a face lift despite heroic efforts at repositioning descended facial fat (Fig. 5).

FACIAL ANALYSIS—THE ROLE OF SKELETAL SUPPORT IN FORMULATING A SURGICAL TREATMENT PLAN

Facial shape and contour is intuitively evaluated when analyzing a patient for facial rejuvenation. Often the two-dimensional considerations seen in photographs are the easiest aspects of aging to identify, and such factors as nasolabial fold depth, jowl prominence, and cervical contour become the primary objectives to improve appearance in the middle-aged face. While these factors are certainly important considerations in treatment planning, the more subtle three-dimensional qualities of facial shape are equally important to evaluate, and are greatly influenced by underlying skeletal support (14).

In evaluating facial shape during preoperative analysis, some of the major factors that I have found helpful to consider include the following.

1. *Facial width, bizygomatic diameter and malar volume.* The emphasis in face lifting over the last 15 years has focused on malar pad elevation. While malar pad elevation and restoration of malar highlights is an important factor in improving facial shape, it needs to be patient specific. Many patients present preoperatively with wide faces, strong malar eminences, and large malar volume with little evidence of malar fat descent. In these individuals, it is necessary to evaluate preoperatively the degree of malar pad elevation required to improve facial shape. While limited degrees of malar pad elevation can be helpful in patients who present with wide bizygomatic diameters, in general, if the malar volume is significantly enhanced in these types of individuals, the aesthetic effect is to make a wide face appear even wider on the front view postoperatively (Fig. 6).
2. *Facial length and the relative vertical heights of the lower and middle third of the face.* Compared with wide faces, patients who present with vertical maxillary excess often have long, thin faces on the front view. As facial fat descends in middle age, it becomes situated anteriorly and inferiorly in the face, and the face appears even longer with age. Malar pad elevation and enhancing malar volume in these types of patients is usually beneficial. As malar volume is enhanced and bizygomatic diameter is increased, the face appears wider on the front view, detracting from relatively excessive facial length (Fig. 7).
3. *Convexity of the malar region juxtaposed to the concavity of the submalar region.* In youth, facial fat is situated overlying the malar and preparotid region. This malar fullness is juxtaposed to a concavity within the submalar region overlying the buccinator. As patients age, the relationship between the malar and submalar regions changes and with it, facial shape changes. As facial fat descends and facial deflation occurs, there is less volume overlying the malar eminence and an associated increase in fullness in the submalar region. As the aesthetic relationship between the malar and submalar regions becomes modified with time, there is a loss of the angular, tapered configuration in shape noted in youth, and middle-aged faces often appear oval. With greater facial fat descent, and an increase in submalar fullness, older faces appear square shaped (Fig. 8).

Preoperatively, an evaluation of the relationship between the malar and submalar regions on the front view, for me, is an essential component of aesthetic treatment planning. For many patients, a restoration in this relationship by increasing malar highlights and malar volume, in association with a restoration of concavity in the submalar region, becomes a central component in improving facial shape (Figs. 9 and 10).

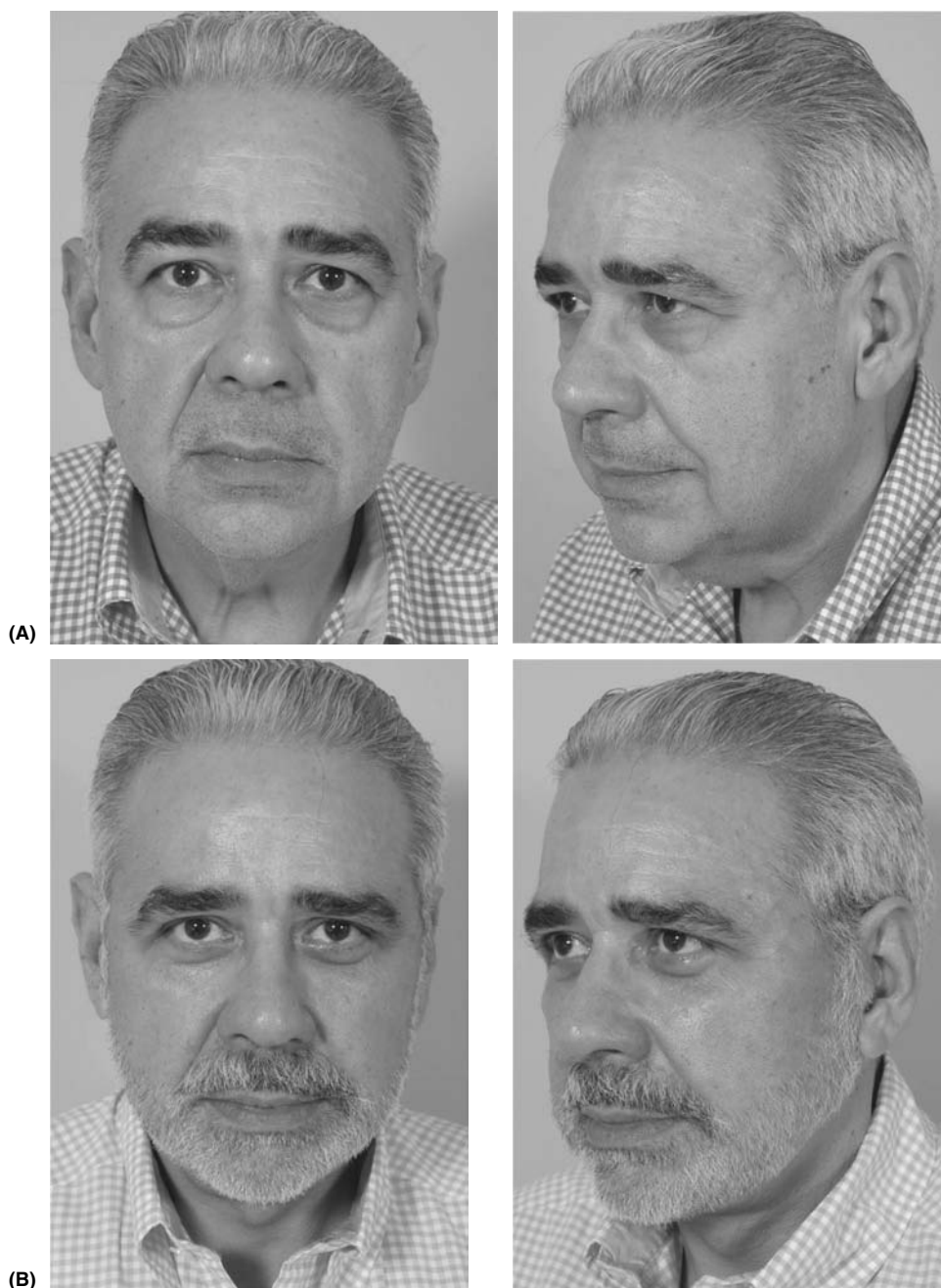


FIGURE 5 (A) Preoperative appearance of a 59-year-old male following a 90-pound weight loss from a gastric bypass procedure. Note the significant areas of facial deflation along the infraorbital rim, lateral orbital rim, and malar region. Also, note the radial expansion of skin and fat lateral to the nasolabial fold, most marked on the right side. Not only does malar fat descend, but attenuation of the retinacular connections between skin, fat, and deep facial fascia lateral to the nasolabial line allows prolapse of soft tissue which accentuates nasolabial prominence. (B) Postoperative result. The areas of deflation along the infraorbital rim, lateral orbital rim, and malar region are improved as facial fat has been repositioned into these regions. The nasolabial folds are somewhat improved following malar pad repositioning, but correction is incomplete, especially on the right. Malar pad elevation helps to flatten the prominent nasolabial fold, but does little to correct radial expansion, with the skin lateral to the nasolabial line remaining prolapsed from its attachments to the facial skeleton. *Source:* From Ref. 14.



FIGURE 6 Facial width and bizygomatic diameter reflect the underlying degree of skeletal support. Patients who exhibit strong malar eminences and wide bizygomatic diameter often benefit from having malar highlights restored, but usually do not require significantly enhancing malar volume (which will cause a wide face to appear wider postoperatively). Shaping considerations in these types of faces usually focus on improving the appearance of the lower two-thirds of the cheek, specifically addressing jowl fat repositioning, as well as creating submalar hollowing which improves the aesthetic relationship between malar and submalar regions. *Source:* From Ref. 14.

The Vertical Height of the Mandibular Ramus and the Horizontal Length of the Mandibular Body

The vertical height of the mandibular ramus and the horizontal length of the mandibular body provide skeletal support for the lower two-thirds of the face. Patients who present with a normal mandibular ramus height, as well as adequate horizontal length of the mandibular body, usually have excellent skeletal support for soft tissue repositioning and are, therefore, less of a surgical challenge. In contradistinction, patients with a short mandibular ramus, an open mandibular plane angle and a short length of the mandibular body typically have poor skeletal support for midface and perioral soft tissue repositioning. These patients are a greater surgical challenge in restoring facial shape, and often benefit from volumetric augmentation, either alloplastic or autogenous, to enhance skeletal support (Fig. 11).



FIGURE 7 Long, thin faces often benefit from an enhancement of malar volume. Superficial musculoaponeurotic system dissection and facial fat repositioning carried anteriorly over the zygomatic eminence allows the surgeon to restore malar volume, thereby increasing bizygomatic diameter. When malar volume is enhanced, the face appears wider, detracting from the relatively excessive facial length. *Left: Before. Right: After.* Source: From Ref. 14.

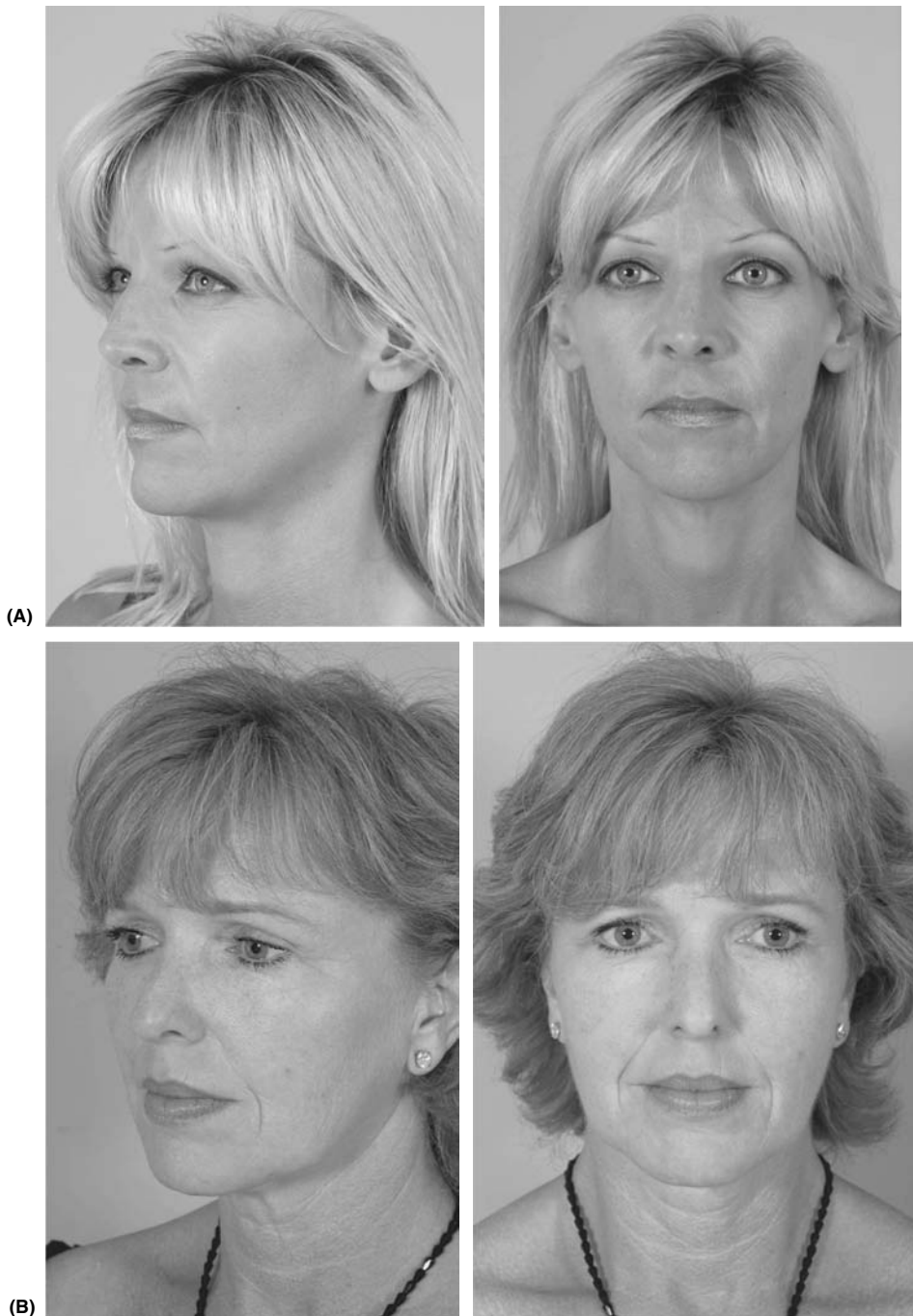


FIGURE 8 (A) This patient demonstrates that the aesthetic relationship between the malar and submalar regions has a significant impact on facial shape. Youthful faces with a tapered configuration typically exhibit a convexity or fullness in the malar region juxtaposed to submalar hollowing. (B) Another patient with similar skeletal support photographed at the age of 51 years. In middle age, facial fat both descends and deflates, accounting for a change in the relationship between the malar and submalar regions. Typically, there is a loss of volume in the malar area associated with an increase in fullness within the submalar region, blunting the relationship noted in youth. Facial shape becomes less tapered, squarer in configuration, with less differential in highlights between the malar eminence and submalar fat

Source: From Ref. 14.



FIGURE 9 (A) Preoperative appearance. Note that facial shape is oval, secondary to malar deflation associated with an increase in submalar fullness. (B) Postoperatively, following malar pad elevation, malar volume is enhanced in association with a restoration of submalar concavity, producing a more angular appearance to facial shape. *Source:* From Ref. 14.

OPERATIVE TECHNIQUE

Surgical Technique—Extended SMAS Dissection

With enough facial laxity to justify a face lift, most individuals will benefit from tightening of the superficial fascial layer as part of the operative plan for facial rejuvenation. Restoration of support to the underlying deeper facial soft tissues has become the key ingredient to my approach to improve facial aging. If the SMAS is thin and tenuous, plication of this layer is an alternative to formal SMAS elevation. Nonetheless, in my opinion, better contouring and longer lasting results are obtained following a formal dissection of the superficial fascia.

In skin flap dissection, it is important to develop uniform skin flaps during the subcutaneous undermining, with care to leave some fat intact along the superficial surface of the SMAS, especially in the regions where the SMAS is to be dissected. If the skin flaps are dissected so that no fat is left along the superficial surface of the SMAS, then the SMAS becomes more difficult to raise, appearing thin, tenuous, and prone to tearing. In an SMAS-type face lift, much of the contouring that is obtained is due to elevation and fixation of the SMAS layer. The more substantial the SMAS flap, often the better long-term results that can be obtained in terms of facial contouring.

I carry the subcutaneous skin flap dissection well into the malar region, and usually the skin overlying the lateral two-thirds of the zygomatic eminence is undermined. I prefer to stop the skin undermining several centimeters lateral to the nasolabial fold rather than undermining the skin to this facial landmark. The reason is that if one limits the dissection of the skin flap in the medial aspect of the cheek, this will preserve some of the attachments that go from SMAS to facial skin. The preservation of these attachments, followed by adequate undermining of the superficial facial fascia (SMAS), will allow the surgeon to reelevate



FIGURE 10 (A) Preoperatively, this patient shows a similar blunting of the relationship between malar and submalar regions. (B) Postoperatively, enhancing malar volume (and bizygomatic diameter) and restoring the concavity within the submalar region makes the face appear more angular in shape, as well as appear vertically shorter. *Source:* From Ref. 14.

anteriorly displaced fat and skin through SMAS rotation rather than to redrape the superficial fascia completely independent of skin flap redraping (3,8).

SMAS Elevation

The dissection of the superficial fascia allows the surgeon to reposition descended cheek fat into the upper lateral face and restore the volumetric highlights overlying the lateral skeletal pillars. As fat is repositioned to these regions, the face appears more structurally supported, assuming a more youthful facial configuration.

The incisions for extended SMAS dissection begin approximately 1 cm inferior to the zygomatic arch to ensure frontal branch preservation. This horizontal incision is continued several centimeters forward to the region where the zygomatic arch joins the body of the zygoma. At this point, the malar extension of the SMAS dissection begins with the incision angling superiorly over the malar eminence toward the lateral canthus for a distance of 3 to 4 cm. On reaching the edge of the subcutaneous skin flap in the region of the lateral orbit, the incision is carried inferiorly at a 90° angle toward the superior aspect of the nasolabial fold. A vertical incision is designed along the preauricular region, extending along the posterior border of the platysma to a point 5 to 6 cm below the mandibular border. In essence, the malar extension of the SMAS dissection simply represents an extension of a standard SMAS dissection into the malar region in an attempt to obtain a more complete form of deep layer support (Fig. 12) (3,8).

The SMAS in the malar region is then elevated in continuity with the SMAS of the cheek. When elevating this flap, the fibers of the orbicularis oculi, as well as the zygomaticus major and minor, are usually evident and the flap is elevated directly along the superficial surface of these muscles. It is important to carry the dissection directly external to these muscle



FIGURE 11 (A) The underlying skeletal support from the mandible has a significant impact on the surgeon's ability to restore facial shape. Patients with a normal height to the mandibular ramus and a long mandibular body are favorable surgical candidates, as there is a significant skeletal support to aid in shaping through facial fat repositioning. (B) In patients with a retrusive mandible, a short mandibular ramus associated with an open mandibular plane angle, there is less skeletal support to aid in facial shaping. These patients represent a greater surgical challenge and often benefit from volumetric augmentation. *Source:* From Ref. 14.

fibers, where a natural plane exists, remembering that the facial nerve branches lie deep to these muscular bellies. The malar SMAS is then elevated until the flap is freed from the underlying zygomatic prominence. Freeing of the SMAS completely from the zygomatic attachments is an important technical point in obtaining the mobility necessary to reposition the malar soft tissue superiorly. To obtain this mobility usually also requires a division of the upper fibers of the masseteric cutaneous ligaments, which will expose the underlying body of the buccal fat pad. The cheek portion of the SMAS dissection is performed beginning directly overlying the parotid gland and then extending this dissection anterior to the parotid utilizing a combination of sharp and blunt dissection toward the anterior border of the masseter (Fig. 13).

In most patients, following extended SMAS dissection of the cheek and malar regions, mobility of the soft tissues lying lateral to the nasolabial fold remain restricted unless the dissection is carried more medially. This restriction in movement results from the undivided retaining ligaments that originate medial to the zygomaticus minor. To improve mobility, I commonly continue malar pad elevation medially in an area where I have not subcutaneously undermined the skin. This dissection is carried directly in the plane between the malar fat and the superficial surface of the elevators of the upper lip. It is usually quite easy to delineate this level of dissection after the malar SMAS elevation is complete, and the superficial surface of the elevators of the upper lip visualized. The scissors are then inserted directly superficial to the elevators of the upper lip and blunt dissection is quickly performed by pushing the scissors in a series of passes bluntly toward the nasolabial fold. I find that when we insert the scissors in the proper plane, the dissection quickly glides through the malar soft tissues and I usually will feel a "snap" as we dissect through the remaining retaining ligaments. Once these structures are divided, one notes greater mobility when traction is applied to the malar portion of the SMAS flap, translating into greater movement along the uppermost portion of the nasolabial fold.

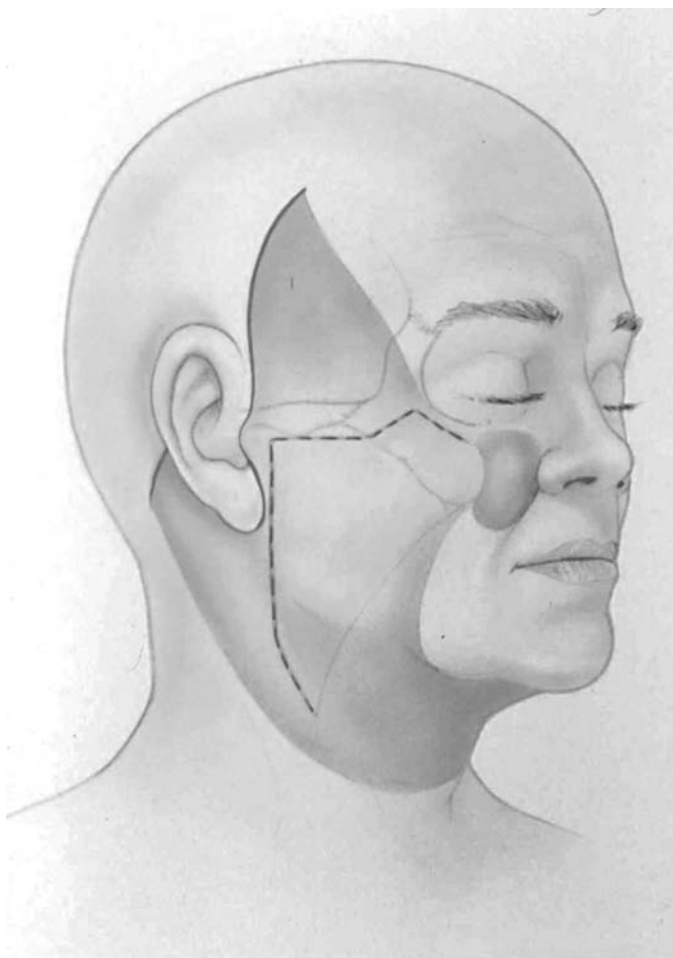


FIGURE 12 The incision design for the extended superficial musculoaponeurotic system (SMAS) dissection is an extension of a standard lateral SMAS dissection. The malar aspect of the dissection allows for the repositioning of facial fat along the infraorbital and lateral orbital rim, as well as restoring malar volume. The SMAS dissection just lateral to the zygomatic eminence provides access to the masseteric ligaments to allow jowl fat elevation and restoring submalar concavity. The dissection laterally in the cheek frees the SMAS from the parotid, providing for repositioning descended platysma, thereby improving mandibular border contour.

AESTHETIC CONSIDERATIONS IN SMAS REPOSITIONING TO IMPROVE FACIAL SHAPE

The aesthetic effects of SMAS repositioning are influenced by three factors: the degree of release, the vector of fat repositioning, and how the superficial fascia is fixated (18). As postoperative contour is dependent on each of these factors, preoperative planning needs to be patient specific in terms of the degree of SMAS release required, the vectors in which facial fat is repositioned, and the location and method for SMAS fixation.

Release

The incision design of an extended SMAS dissection allows for complete release of the SMAS from its underlying retaining ligamentous attachment in the lateral midface. As surgeons, there is a tendency to believe that a greater degree of SMAS dissection equates with a better result, but this has not been my experience. Rather, precision in the degree of SMAS dissection and its release from the retaining ligaments as dictated by the aesthetic needs of a patient increases surgical control and consistency while minimizing morbidity (14).



FIGURE 13 Illustration showing the typical extent of superficial musculoaponeurotic system (SMAS) dissection in the malar region, cheek, and neck. The end point in SMAS dissection is to obtain adequate mobility with redraping. This requires freeing the SMAS in the malar region completely from its zygomatic attachments. To reelevator jowl fat back upward into the cheek usually requires division of both the zygomatic ligaments and the upper portion of the masseteric cutaneous ligaments. The SMAS in the cheek below the parotid duct usually is elevated to the anterior border of the masseter, and occasionally, division of the lower masseteric cutaneous fibers is required. The SMAS in the neck is elevated off the tail of the gland, extending anteriorly within the areolar plane deep to the platysma, until adequate mobility is obtained in contouring the submandibular and submental regions of the neck. The key to SMAS elevation is obtaining adequate mobilization that allows reelevation of facial fat without undue tension. *Source:* From Ref. 17.

How much to release the SMAS, and how high and anterior to carry the SMAS dissection, needs to be decided preoperatively. In patients who present with adequate malar volume, wide bizygomatic diameter and little evidence of malar pad descent, it is usually unnecessary to carry the SMAS dissection medial to the lateral orbital rim (although I usually carry the dissection high within the malar eminence to allow fat repositioning along the infraorbital and lateral orbital rims). The region where the malar eminence structurally curves from its anterior projection toward its lateral extension with the zygomatic arch is a useful landmark in planning the location and degree of SMAS release. In patients with a wide bizygomatic diameter, limiting the SMAS dissection to the lateral aspect of the malar eminence will not increase facial width on the front view. Typically, the shaping considerations for patients with a wide face are focused on reducing fullness in the submalar region. Sub-SMAS dissection along the lateral aspect of the zygomatic eminence provides exposure to the juxtaposed masseteric ligaments, allowing the submalar fat to be repositioned superiorly along the concavity of the underlying buccinator, thereby restoring submalar hollowing (Fig. 14).



FIGURE 14 Patients with wide bizygomatic diameters and good underlying skeletal support typically do not require a significant anterior malar dissection to improve facial shape. Most commonly, the superficial musculoaponeurotic system dissection in these patients (while kept high) is extended only as medial as the lateral orbital rim, so that malar volume restoration is limited to the lateral aspect of the zygomatic eminence. The shaping considerations for these types of faces usually emphasize reducing fullness within the submalar area, as well as jowl fat elevation. Note that postoperatively the patient's face appears more tapered and thinner in morphology through facial fat repositioning without removal of facial fat. *Left: Before. Right: After.* Source: From Ref. 14.

Vertically long faces often benefit from carrying the malar portion of the extended SMAS dissection anteriorly and medially to the lateral orbital rim and onto the anterior surface of the malar eminence to increase anterior malar volume. Carrying the SMAS dissection more medially allows the surgeon to enhance anterior malar volume and restore malar highlights anteriorly over the zygomatic eminence, thereby increasing facial width on the front view (Fig. 15).

Vectors of Fat Elevation—Facial Asymmetry

All patients exhibit some degree of facial asymmetry. Commonly, one side of the face is vertically longer and the short side of the face is usually wider than the long side. Malar highlights are typically more superiorly located on the long side of the face and, with age, facial fat tends to descend in a more vertical direction on the long side. As facial asymmetry and facial skeletal configuration are asymmetric in most individuals, it follows that the vectors of fat elevation (SMAS repositioning) should be specific for the right and the left side of the face.

The vector in which the SMAS is repositioned has a significant impact on the location and volume of elevated facial fat, thereby influencing facial shape. Decisions regarding the direction of SMAS vectoring for the right and left side of the face are best determined preoperatively, as it is very difficult to make aesthetic vector judgments intraoperatively with the patient recumbent.

SMAS vectors influence postoperative facial shape. Vertical SMAS repositioning typically provides a larger amount of fat for malar eminence enhancement, as well as allowing for



FIGURE 15 Faces that are more dominated by their facial length (especially the lower third of the face) usually benefit from malar volume restoration. To enhance malar volume requires the superficial musculoaponeurotic system dissection be carried toward the anterior aspect of the zygomatic eminence, such that malar volume is increased in this region. *Left: Before. Right: After.* Source: From Ref. 14.

a reduction in fullness within the submalar region as fat is forced vertically along the concavity of the buccinator. For this reason, vertical SMAS vectors are often indicated to reshape round, full faces, allowing them to appear more tapered and thinner postoperatively (Fig. 16). If the SMAS is vectored more obliquely, there is less volume of fat brought into the malar region and a greater volume of fat repositioned into the submalar region. Oblique SMAS repositioning is therefore helpful in elderly patients who appear gaunt over the buccal recess, as it allows the surgeon to volumetrically enhance the submalar region (Fig. 17) (14).

SMAS Fixation

In a two-layer SMAS-type face lift, the tension of contouring is placed on the superficial fascia rather than the skin envelope. For this reason, the fascial quality and tensile strength of the superficial fascia has an influence on both the longevity of result and the volume of fat, which can be repositioned intraoperatively and maintained postoperatively. In other words, soft tissue quality influences long-term contour, and is the primary reason why face lifts in young patients are more predictable.

In an effort to improve fascial quality in an SMAS-type face lift, for the last 10 years, I have incorporated vicryl mesh into the SMAS fixation (11). It is my observation that incorporating vicryl mesh into SMAS fixation not only improves the longevity of these procedures, but provides the surgeon greater aesthetic control in terms of restoring facial shape. Incorporating the vicryl mesh allows the surgeon to secure the SMAS under greater tension (while preventing the sutures from “cheese wiring” through the SMAS), allowing the surgeon to reposition a greater volume of fat into specific regions of the midface. Greater control in fixating repositioned fat provides greater control in restoring facial shape.



FIGURE 16 Vectors of superficial musculoaponeurotic system (SMAS) elevation have a significant impact on facial shape. Vertical repositioning of the SMAS allows the surgeon to enhance malar volume and reduce fullness within the submalar region, as fat is forced up along the concavity of the buccinator. Restoration of submalar hollowing through SMAS vectoring is useful in contouring full faces, making them appear thinner postoperatively. In this patient, a small amount of jowl defatting through needle aspiration was also performed. *Left: Before. Right: After.* Source: From Ref. 14.

If malar volume requires enhancement, once the SMAS is initially fixated, adding a few more sutures between the SMAS flap and the periosteum of the malar eminence allows the surgeon to imbricate more malar fat superiorly over the zygoma, thereby increasing malar volume. Essentially, this type of stacking fixation imbricates malar fat onto both mesh and zygomatic eminence, aesthetically resulting in an increase in bizygomatic diameter (Fig. 18).

If the predominant aesthetic problem is to reduce fullness within the submalar region, SMAS release of the upper masseteric ligaments and vertically vectoring the SMAS tends to increase submalar hollowing. Adding fixation sutures just lateral to the malar eminence also accentuates submalar concavity. This method of SMAS vectoring and fixation is useful in shaping round, full faces, without having to overly rely on the removal of facial fat to improve facial shape.

Suturing in the preparotid region repositions descended platysma muscle superiorly over the mandibular border, thereby accentuating mandibular border and cervical contour. Adding sutures into the SMAS fixation along the preparotid region is also useful in providing volumetric enhancement laterally in the cheek, improving an aesthetic subunit which typically deflates with age.

THE AESTHETIC EFFECT OF STRONG VERTICAL SKIN TENSION ON FACIAL SHAPE

Strong, vertical shifting of the cervicofacial flap has traditionally been a maneuver utilized in many face lift techniques. While skin envelope tightening can produce a dramatic effect in terms of the improvement of facial laxity, the aesthetic effects of vertical skin shift on facial shape have been poorly delineated. Specifically, when skin is shifted in a cephalad direction, the effect of skin tension commonly produces a depression or flatness in the preparotid



FIGURE 17 This patient exhibits asymmetry in the submalar region preoperatively. Note that she appears hollow and concave on the right, while she is fuller on the left side. For this reason, the superficial musculoaponeurotic system was vectored obliquely on the right to volumetrically enhance the submalar region, while it was vertically vectored on the left side to restore submalar hollowing and balance the two sides of her face. *Left: Before. Right: After.* Source: From Ref. 14.

region, an area that typically deflates with aging. If the preparotid region remains deflated despite fat repositioning, vertical skin vectoring will further depress and flatten this region. In my opinion, vertical skin shift all too often produces an unnatural tightness to facial shape, producing the typical stigma associated with rhytidectomy (Fig. 19) (11,14). If the surgeon has been successful in enhancing contour through repositioning descended facial fat, the use of strong vertical skin tension is neither desirable nor necessary to enhance the postoperative result.

The primary aesthetic advantage of performing skin undermining separately from SMAS dissection is that it allows facial fat repositioning to be independent of skin flap redraping, and the skin envelope tension is not required to improve facial shape. This tends to produce a more natural aesthetic result.

CONCLUSION

Thirty years after the milestone work of Mitz and Peyronie (2), plastic surgeons continue to struggle in their quest for a perfect solution to rejuvenate the aging face (18–38). Over this time period, the surgical spectrum has swung from wide skin and SMAS dissection to short scar face lifts with imbrication or SMAS resection, in an attempt to limit morbidity while seeking to improve consistency in results (31,38).

In my opinion, surgical rejuvenation of the aging face suffers not from technical solutions, but rather from a uniform method of preoperative analysis. Unlike rhinoplasty, where preoperative analysis is defined by angles and measurements of length and projection, aging faces vary greatly in terms of skeletal support, volume of fat, fascial quality, and skin quality. The variability of facial shape from patient to patient, as well as how individual patients

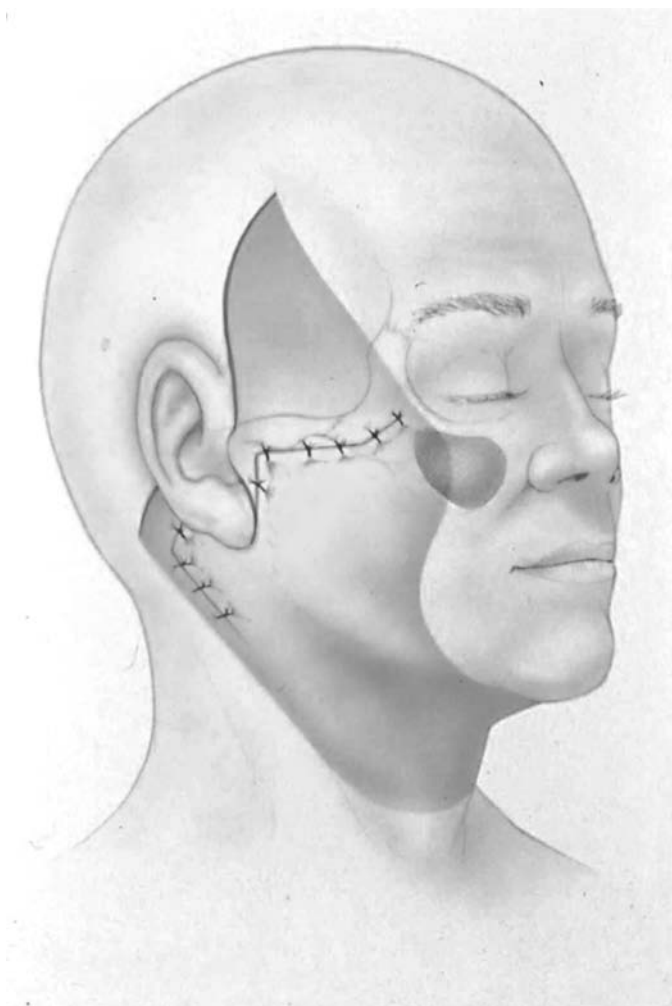


FIGURE 18 Fixation of the superficial fascia affects facial shape. The advantages of incorporating vicryl mesh into the superficial musculoaponeurotic system (SMAS) fixation is that it provides the surgeon greater control in determining the position and volume of repositioned fat. Suturing in specific areas along the SMAS incision affects volume for a particular region. Adding sutures in the malar region allows the surgeon to bring a greater amount of malar fat onto the zygomatic eminence, thereby enhancing malar volume. Adding sutures in the region just lateral to the zygomatic eminence forces cheek fat superiorly along the concavity of the buccinator, thereby reducing submalar fullness. Sutures placed in the preparotid region force descended platysma superiorly over the mandibular border, improving mandibular border contour. *Source:* From Ref. 14.

age, makes a uniform system of analysis difficult. Artistic vision and artistic goals, as to what will aesthetically improve for a particular patient, is also surgeon specific.

From my perspective, independent of the technique, the primary goal in face lifting is to improve facial appearance while minimizing signs that a surgical procedure has been performed. Understanding the nuances of superficial fascia repositioning (repositioning facial fat), whether it be utilizing imbrication, SMAS resection, or formal SMAS flap elevation, provides the surgeon only with a mechanism to restore facial shape. What is critical to the end result is preoperative analysis and accurately delineating the aesthetic goals prior to surgery. Formulating a treatment plan to restore facial shape which is patient specific, and is based on the volumetric requirements as influenced by underlying skeleton support is the key element to increase consistency in face lifting. The technical solution thereby becomes subservient to



FIGURE 19 The aesthetic consequences of vertical redraping of the cervicofacial skin flap are much different than vertical superficial musculoaponeurotic system redraping. Strong vertical shift of the skin flap produces not only temporal hairline distortion, but also accentuates flatness in the prepatrid region (which typically deflates with age). Lateral flatness can produce a tight, unnatural appearance all too commonly associated with patients having undergone a rhytidectomy. *Source:* From Ref. 14.

the aesthetic destination. Good surgeons can get good results through a variety of techniques as long as there is an understanding on how to vary technique according to the aesthetic needs of the patient.

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6 Nonsyndromic Craniosynostosis: Current Treatment Options

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INTRODUCTION

The significance and etiology of abnormal skull shape has been under investigation since ancient times. Its pathophysiology was first described in 1851 by Virchow, who recognized that “bony expansion ceases in a direction perpendicular to the synostosed suture, with compensatory expansion in the opposite direction” (1). The premature closure of calvarial sutures is referred to as craniosynostosis and occurs at an estimated frequency of 0.4/1000 births (2). Nonsyndromic, or isolated, craniosynostosis predominates and is defined as suture fusion that creates functional impairments related to local effects of the fusion.

The first surgical repair of craniosynostosis was performed by Lannelongue in 1892 (3). Mohr et al. published their experience in craniosynostosis repair emphasizing the etiologic involvement of the anterior skull base, and employing extensive vault reconstruction (4). In 1978, Jane et al. published their π technique to accomplish immediate reconstruction of frontal bossing (5). Anderson, in 1981, described his experience in the treatment of coronal and metopic synostosis (6). Then in 1984, Jane et al. published their technique for the treatment of unilateral coronal synostosis, which included dural plication (7). In 1985, Albright described his correction of sagittal synostosis, which included extensive occipital circular, and parietal wedge craniectomies (8). Delashaw et al., in 1986, reported their technique for the correction of metopic synostosis also implementing dural plication (9). Greene and Winston reported correction of scaphocephaly with sagittal craniectomy and biparietal morcellation (10). In 1990, Persing et al. published their work using near-total cranial vault reconstruction in concert with fronto-orbital advancement to correct bilateral coronal synostosis (11). Cohen et al., in 1991, utilized an advancement-onlay technique for fronto-orbital remodeling in craniosynostosis repair (12). In 1991, Shaffrey et al. published their technique in extensive reconstruction in metopic synostosis repair (13). Jimenez and Barone, in 1995, published their “Sunrise” technique in the correction of occipital plagiocephaly (14). Clearly, craniosynostosis repair has gone through vast evolution from simple “suturectomy” to extensive cranial vault reconstruction.

Quantifying the efficacy of surgical repair has been problematic. Cohen et al., in 1994, published an attempt to objectively assess the aesthetic result in metopic synostosis repair (15). A number of papers have used reoperation rates as an indirect measure of surgical outcomes in single suture craniosynostosis repair. McCarthy et al. reviewed the experience at New York University Medical Center in 1995 and found a reoperation rate of 13% (16). In reviewing 167

consecutive patients undergoing open correction of single suture synostosis, Williams et al. found a reoperation rate of 5.9% (17).

The era of *minimally invasive* craniosynostosis repair, implementing endoscopic visualization, began with the work of Vicari in 1994 (18). Barone and Jimenez reported their technique utilizing endoscopic strip craniectomy combined with postoperative helmet molding in 1999 (19). Jimenez et al. published their assessment of safety, efficacy, and results of their endoscopic-assisted strip craniectomy and postoperative cranial orthotic molding in 2002 (20). Subsequently, Cohen et al. published a technique describing cranial osteotomies performed through small scalp incisions with endoscopic guidance sometimes utilizing bioresorbable plates (21).

The purpose of this article is to present our current approach to patients with nonsyndromic craniosynostosis, outlining the place of both open, conventional approaches and newer, minimally invasive, endoscopic assisted craniosynostosis correction.

CURRENT APPROACHES

Pediatricians in our region are encouraged to identify and refer all infants with abnormalities in head shape as early as possible to insure a prompt diagnosis of patients with craniosynostosis. Patients are routinely examined by the pediatric neurosurgeon and craniofacial surgeon. Frequently, patients are examined by a pediatric neuroophthalmologist and geneticist. Team care is available for all patients with complex associated anomalies and/or syndromic craniosynostosis.

The Open Technique

Patient Preparation

In children undergoing simple sagittal or lambdoid synostectomies, two intravenous (IV) lines are used, but a Foley catheter and intra-arterial line are not routinely inserted. Simple sagittal and lambdoid synostectomies are performed with the patient in prone position on a horseshoe headrest. In patients undergoing either a Pi procedure (5) or a major cranial vault reconstruction for sagittal synostosis, the modified prone position with the neck hyperextended is used. For patients with metopic, unicoronal, and bicoronal synostosis, the patient is operated on in the supine position. The incision is marked with a pen and generally performed in a wavy S-shaped fashion behind the hairline. A Colorado needle-tip electrocautery is used for the skin incision. The dissection is performed in the subgaleal plane. This is carried out down to the level of the supraorbital rims, and then the periosteum is incised along the insertions of the temporal muscles and then horizontally at the level of the anterior fontanelle. This permits subperiosteal elevation of a generous tongue of periosteum and galea. Subperiosteal dissection is then performed to expose the supraorbital rim and lateral orbital rim down to the body of the zygoma, and intraorbitally to the level of the inferior orbital foramen. In patients undergoing correction of metopic, unicoronal, and bicoronal synostosis, a bifrontal craniotomy is always carried out. The anterior cranial fossa is then exposed to perform the fronto-orbital osteotomies. All children undergoing correction of craniosynostosis at our institution are typed and crossed for either donor-directed or banked blood. Before beginning the surgical procedure, the blood is transported to the refrigerator in the operating room, where it is immediately available at the commencement of the case.

Sagittal Synostosis

Depending on the degree of deformity and the age of the patient, a variety of operations have been used for correction of sagittal synostosis. In young infants with mild scaphocephaly, a simple sagittal synostectomy is carried out. For children who present with moderate frontal prominence without saddling deformities or significant occipital abnormalities, a Pi procedure (20) is used. A major cranial vault reconstruction in an infant is reserved for children with severe skull deformities.

Metopic Synostosis

Although some variation of technique is used depending on the degree of deformity, in general the surgical approach is fairly routine for correction of metopic synostosis (22). After frontal craniotomy, bilateral supraorbital rim osteotomies are performed without a temporal tenon or Z-plasty unless wires are being used for stabilization. In patients with severe hypotelorism, correction may be incomplete with the above technique. In such cases, we have used a modified medial orbital osteotomy to increase the degree of interorbital separation. In these cases, the osteotomies are performed in situ.

Unicoronal Synostosis

The surgical approach to unicoronal synostosis again depends on the degree of deformity. Again, after a bifrontal craniotomy, an osteotomy of both supraorbital rims is carried out. The entire complex is then removed. In children with malar recession, the osteotomy along the lateral orbital rim may be carried into the zygoma and even include the inferior orbital rim. In cases in which the nasal deviation is severe, the osteotomy may be performed low across the nasal dorsum. All osteotomies are stabilized with resorbable plates and screws.

Bicoronal Synostosis

Again, a frontal craniotomy that includes the fused coronal sutures is performed and a bilateral supraorbital rim osteotomy is carried out. Fronto-orbital advancement and cranial reshaping are performed and the bones are stabilized with rigid, resorbable fixation.

The Endoscopic, Minimally Invasive Technique

We are careful in the selection of patients who are candidates for the endoscopic approach. Most importantly, the patient needs to be under four months of age if possible. In patients with mild sagittal and metopic synostosis, we are able to operate on patients as old as six months. In patients with mild sagittal synostosis, we have performed the endoscopic procedure in children as old as nine months.

Our standard approach to mild sagittal synostosis is simple strip craniectomy carried out endoscopically. Children are placed in a helmet or band within 7 to 10 days after surgery. In patients with moderate to severe sagittal synostosis, more extensive wedge osteotomies and radial osteotomies may be necessary and in some patients immediate correction is carried out with stabilization of the bony segments with resorbable plates and screws. Orthotic devices are usually necessary for three months. Some children will require a second helmet or band.

In patients with mild to moderate metopic synostosis, strip craniectomy with or without additional osteotomies is performed. Again, the helmet or band is placed within 7 to 10 days if possible. Orthotic treatment usually takes up to six months and most patients require a second helmet or band. For patients with more severe deformities who are under the age of four to six months, the endoscopic assisted fronto-orbital advancement is performed or an open approach is recommended.

Patients with unicoronal synostosis are managed the same as patients with metopic synostosis. Endoscopic assisted fronto-orbital advancement is performed on children with more severe deformities. Helmets or bands may be needed for as long as a year in some cases and second helmets and bands are the rule.

Patient Preparation

Two large IV lines and an arterial line are inserted in the extremities. Patients receive decadron and IV antibiotics for 24 hours as well as mannitol (0.5 g/kg) during the procedure. After induction, the endotracheal tube is secured and the patient is placed in a prone, chin-up position on a beanbag and headrest (sphinx position), for sagittal correction. Patients undergoing metopic or coronal correction are placed in supine position. Doppler is routinely used to monitor for air embolism.

Sagittal Synostosis

W-type incisions are made in the midline approximately 2.5 to 3 cm in transverse length at the anterior fontanelle and the lambdoid suture (Figs. 1 and 2). Skin staples are applied at both ends of the incision to limit tearing of the skin with subsequent retraction. Subgaleal dissection is followed by careful dissection of the dural edge at the anterior fontanelle (Fig. 3). A burr hole is placed on either side of the posterior aspect of the sagittal suture and connected after the underlying sagittal sinus is freed. Subosteal-epidural dissection is then carried out under endoscopic visualization (Fig. 4). Using heavy scissors, the sagittal suture is removed (Figs. 4 and 5). For patients with more severe deformities, lateral wedge osteotomies and radial/frontal and occipital osteotomies are performed to allow biparietal outfracturing and correction of the anterior-posterior and transverse dimensions (Fig. 5). In more severe cases, cranial vault remodeling is maintained with bioresorbable plates and screws to accomplish an immediate reconstruction and shorten the duration and/or eliminate the need for postoperative helmet/band (Fig. 6). Shown in Figure 7 is the resected sagittal suture being delivered through the anterior skin incision. In Figure 8, the endoscopic appearance of the wedge osteotomies, which are created anteriorly and posteriorly on either side of the skull is shown, while in Figure 9, the wedge osteotomy has been closed with gentle pressure and fixated in position with resorbable mesh and screws which, in our present approach, are rarely utilized. Patients are placed in helmets/bands 7 to 10 days after the operation. Representative preoperative and postoperative photographs of a typical patient with sagittal synostosis are shown in Figure 10A and B, respectively.

Presently, patients with mild to moderate sagittal synostosis under six months of age are treated with simple strip craniectomies and/or a combination of wedge and radial osteotomies and osteotomies without fixation. In patients with severe deformity, the more extensive surgery as outlined and illustrated above is routinely performed and resorbable plates and screws are utilized in about 50% of cases.

Metopic Synostosis

For mild to moderate metopic synostosis, simple strip suturectomy with or without bifrontal osteotomies is carried out. Helmets/bands are placed within the first two weeks of surgery. The strip is kept narrow (1 cm) to insure complete bony healing. Helmets/bands are necessary for up to six months. We typically will not offer the endoscopic assisted, minimally invasive approach to patients older than five months.

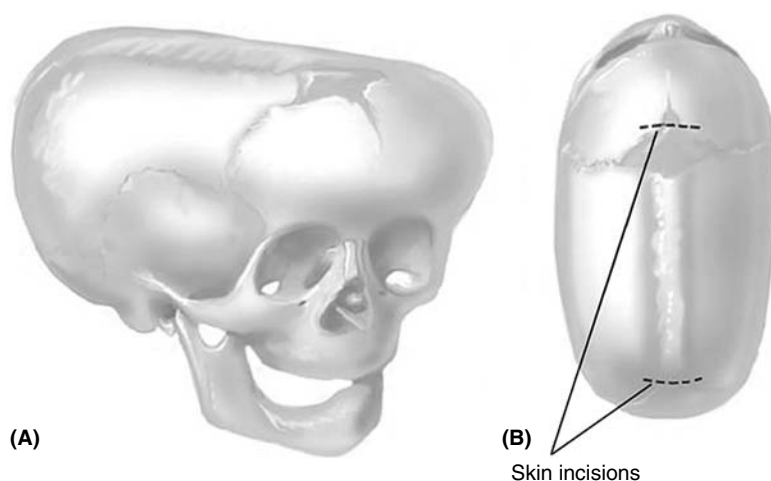


FIGURE 1 (A) Sagittal synostosis with scaphocephaly. (B) Skin incisions. A W-line incision is now preferred for better camouflage in the hair. *Source:* From Ref. 21.

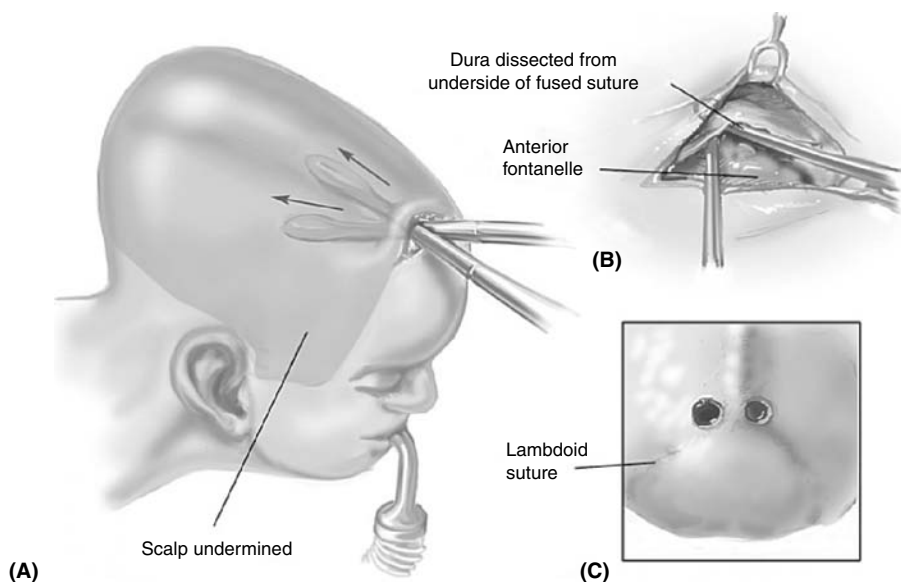


FIGURE 2 Dissection is initiated through the anterior fontanelle and through burr holes at the junction of the sagittal and lambdoid sutures. (A) Scalp undermined. (B) Dura dissected from anterior fontanelle. (C) Posterior burr holes connected and enlarged. *Source:* From Ref. 21.

The proposed osteotomies for endoscopic treatment of the severe metopic suture with attendant trigonocephaly are depicted in Figure 11. A transverse W-style skin incision is made approximately over the anterior fontanelle to access the synostosed metopic suture (Fig. 12). Bilateral upper blepharoplasty incisions expose the supraorbital and lateral orbital rims (Fig. 12). A subgaleal, supraperiosteal dissection exposes the metopic suture. Either through the vestigial anterior fontanelle or via burr holes on either side of the fused metopic suture, endoscopic-assisted dissection frees the underlying sagittal sinus from the bone. Using a rongeur and/or heavy Mayo scissors, a metopic suture osteotomy is carried out down to the nasofrontal junction. The endoscope is used to help prevent and inspect for dural tears.

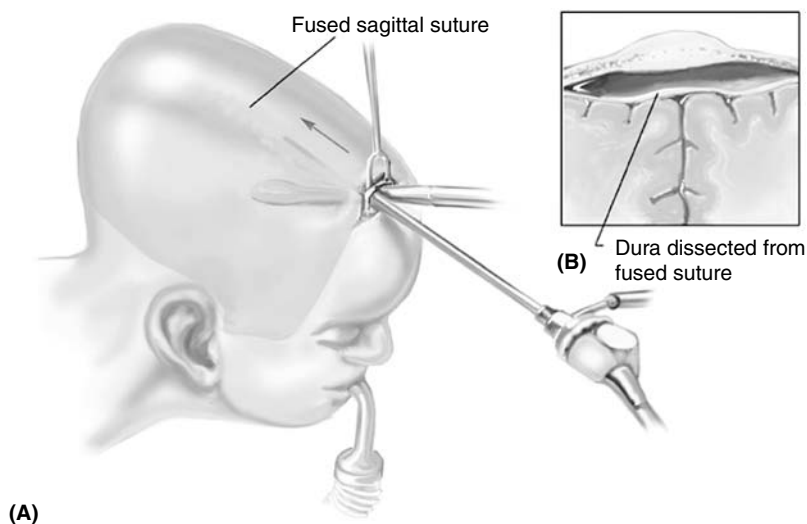


FIGURE 3 (A) The dura being dissected with endoscopic assistance. (B) Dura separated from fused suture. *Source:* From Ref. 21.

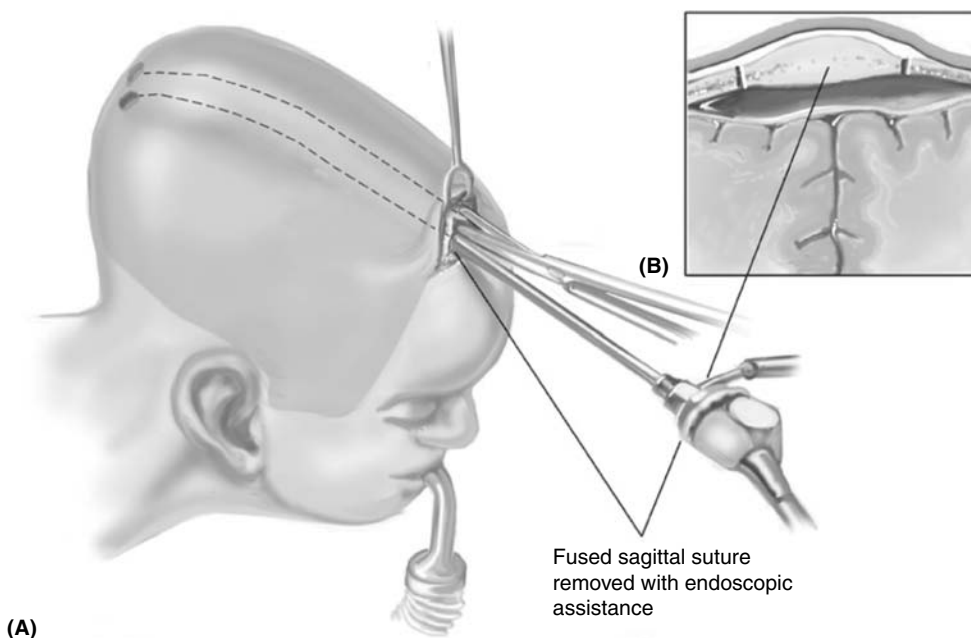


FIGURE 4 The sagittal suture is resected with straight Mayo scissors under endoscopic guidance. (A) Fused suture removed. (B) Osteotomy and removal of fused suture. *Source:* From Ref. 21.

The lateral fronto-orbital osteotomy is depicted in Figure 13. This is performed with curved Mayo scissors down to the lateral orbital rim, which is cut with a reciprocating saw or 3 mm osteotome through the upper eyelid incision. The superior orbital osteotomy is carried out with a reciprocating saw and/or osteotome from the orbital roof inward, protecting the dura and guiding the osteotomy line with endoscopic retraction and visualization (Fig. 14). Having completed the lateral fronto-orbital osteotomies bilaterally and superior orbital osteotomies, gentle digital pressure is used to greenstick fracture the fronto-orbital segments.

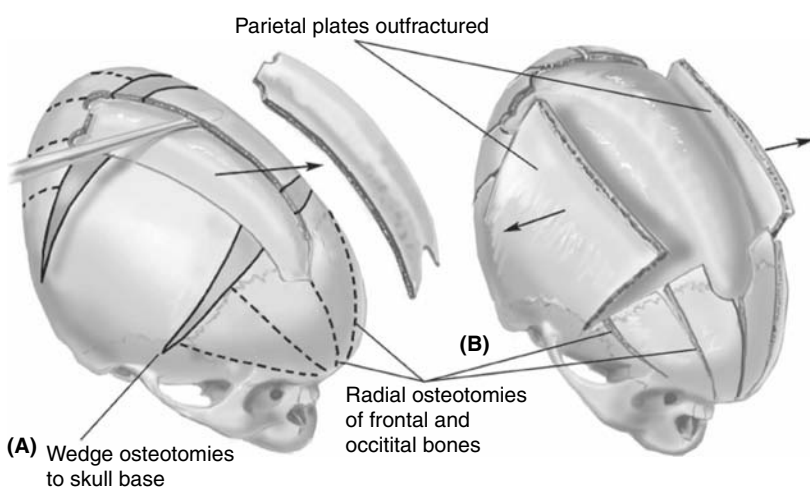


FIGURE 5 Depending on the specific deformity, a variety of osteotomies can be employed. Shown here are wedge osteotomies to allow for anteroposterior shortening and radial osteotomies of the frontal bone to correct bossing. (A) Extensive osteotomies can be made through the small incisions. (B) Sagittal suture removed, bone recontoured. *Source:* From Ref. 21.

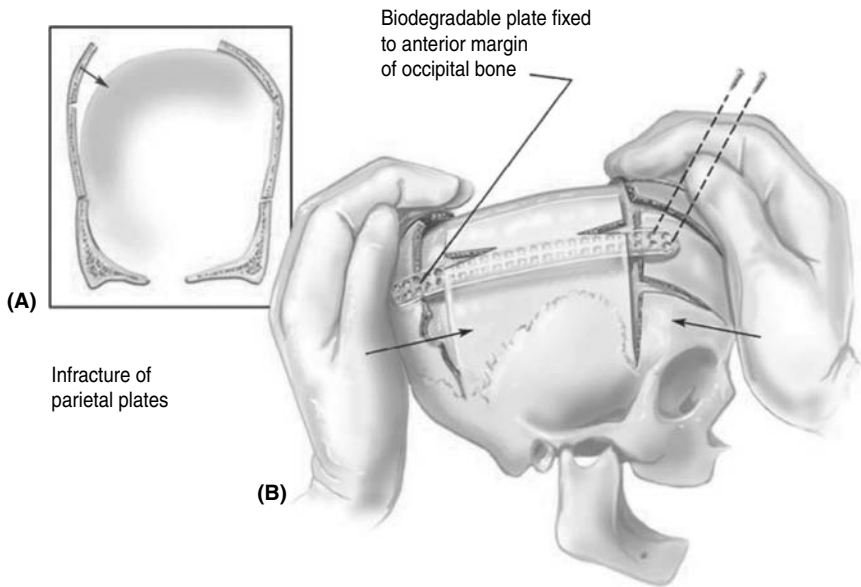


FIGURE 6 Immediate correction and fixation with resorbable plates can be carried out. Shown in this drawing is one of the fixation techniques using a 165 mm resorbable plate. Currently, we use smaller plates fabricated from a sheet of mesh as shown in figure. (A) Infracture of the parietal plates to round out the skull. (B) Immediate anterior posterior shortening with increase in skull width can be performed and stabilized with resorbable plates, although this option is only used in severe cases. *Source:* From Ref. 21.

These segments are then rotated laterally and downward, while simultaneously advancing the superior-lateral orbital rims (Fig. 15) demonstrates the use of bioresorbable plates, which maintain the advancement of the fronto-orbital segments. Bone grafts or resorbable mesh can be cut and placed inside the orbit in such a manner as to maintain the lateral orbital rim



FIGURE 7 Removal of the fused sagittal suture.

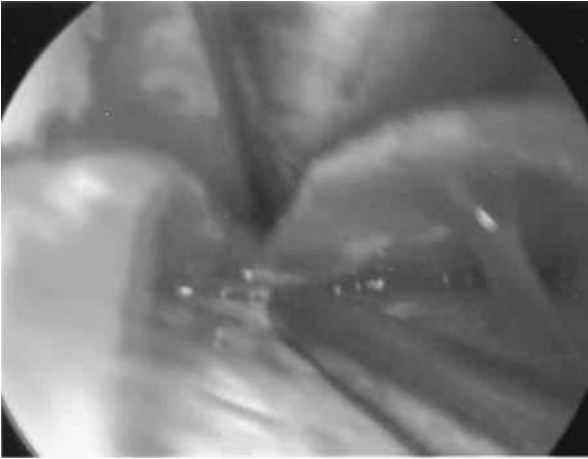


FIGURE 8 Wedge osteotomy. When closed, anteroposterior shortening with horizontal widening of the skull occurs.

advancement, while separating the dura from the orbital contents. Preoperative and two year postoperative photographs are shown in Figures 15B,D and 16A,C, respectively. Patients with mild and moderate trigonocephaly undergo strip craniectomies (1 cm) with or without skull osteotomies and are placed in helmets/bands 7 to 10 days after operation.

Unilateral Coronal Synostosis

For mild to moderate unilateral coronal synostosis, strip suturectomy is carried out. The strip is kept narrow (about 1 cm in width). Helmets/bands are applied within about two weeks of surgery and generally maintained for up to six months. Patients over five months are not considered to be candidates for the endoscopic assisted approach.

The unilateral coronal synostosis endoscopic repair for severe cases is similar to the metopic synostosis correction. A transverse skin incision is made lateral to the anterior



FIGURE 9 Wedge osteotomy closed and resorbable plate applied.

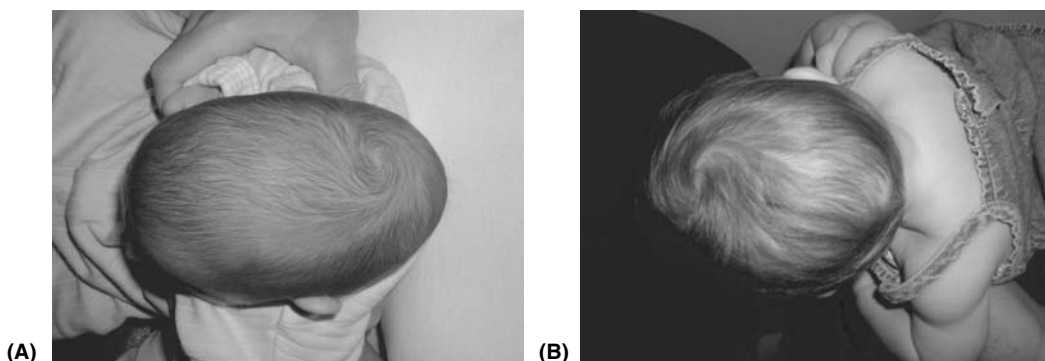


FIGURE 10 (A) Preoperative photograph of four-month-old female with sagittal synostosis. (B) Postoperative photograph three months following endoscopic-assisted correction of sagittal synostosis.

fontanelle on the ipsilateral side of the fused suture. An ipsilateral upper eyelid incision is made as well (Figs. 17 and 18). Careful dissection along the undersurface of the fused coronal suture and frontal bone is carried out with endoscopic assistance and the suture is resected with scissors and/or a rongeur. Only a 1 cm segment which includes the fused coronal suture is removed (Fig. 19). The dura is then separated under endoscopic control from the frontal bone done along the anterior skull base and orbital roof. The lateral fronto-orbital osteotomy followed by the superior orbital osteotomy is performed under endoscopic

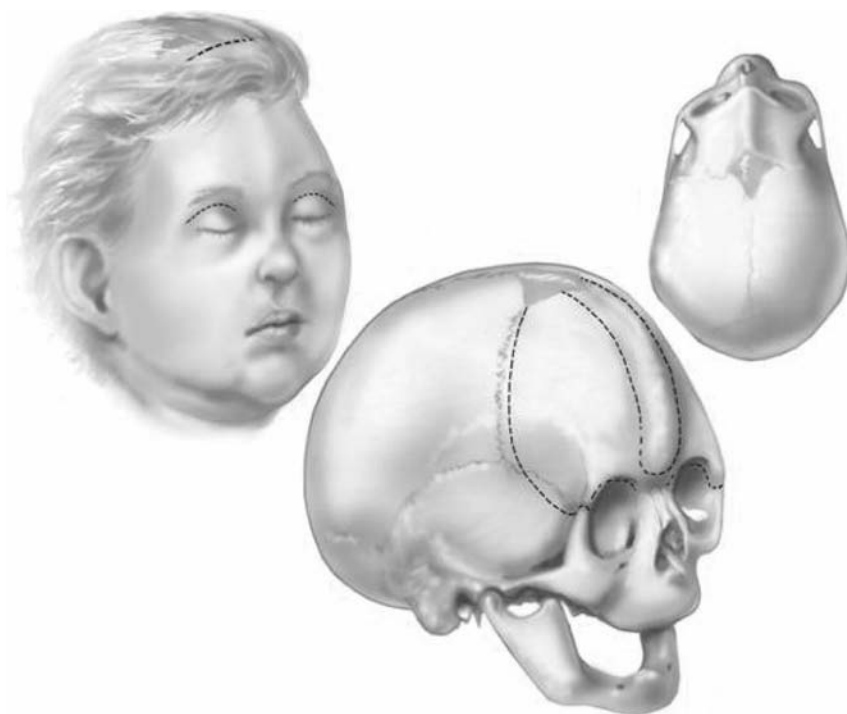


FIGURE 11 The proposed incisions over the anterior fontanelle and both upper eyelids are shown as well as the proposed osteotomy cuts to remove the fused metopic suture and create the fronto-orbital segment. *Source:* From Ref. 23.

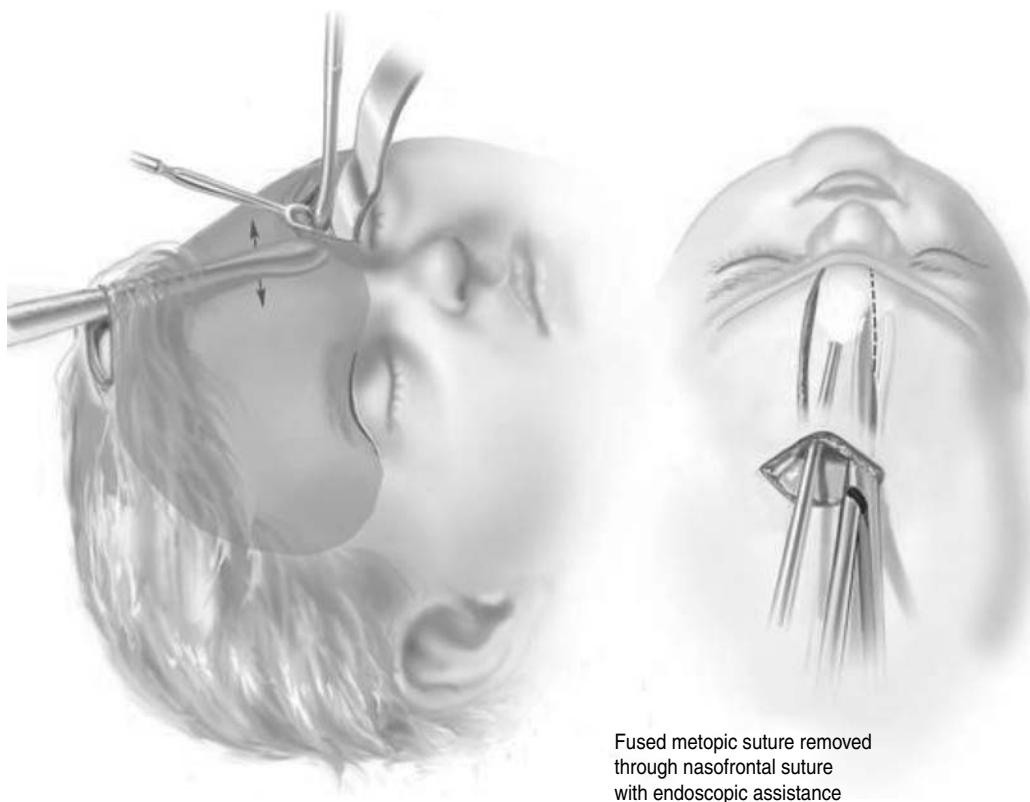


FIGURE 12 With endoscopic guidance, the fused metopic suture is resected. *Source:* From Ref. 23.

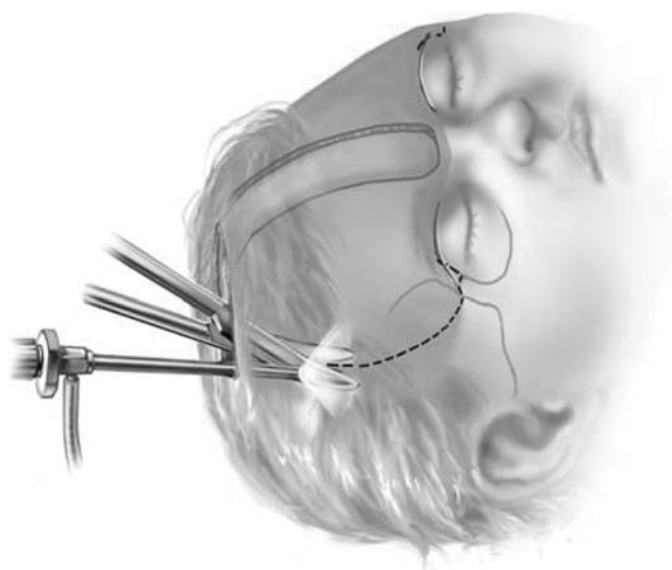


FIGURE 13 An osteotomy in front of the coronal suture is made with scissors down to the lateral orbital rim. *Source:* From Ref. 23.

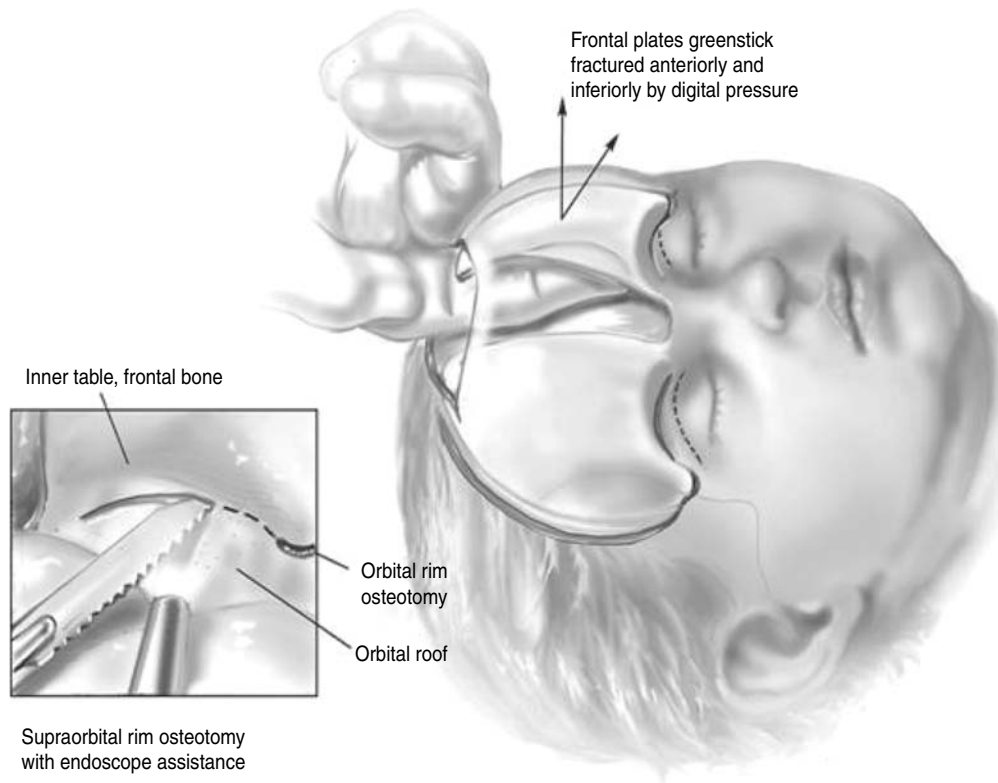


FIGURE 14 A reciprocating saw and/or osteotome is used to make the supraorbital osteotomy. The saw may be introduced through the eyelid or through the frontal osteotomy. Presently, we place the endoscope intracranially and retract the brain, while making an osteotomy across the orbital roof with the saw and/or 3 mm osteotome. *Source:* From Ref. 23.

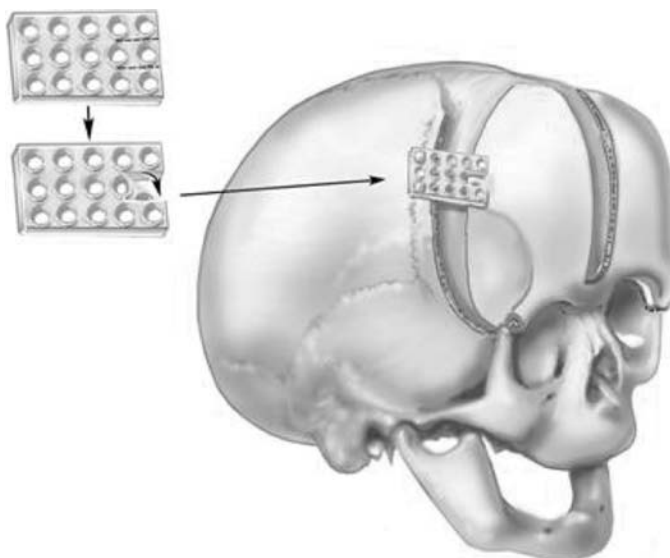


FIGURE 15 Modified mesh to stabilize the advanced fronto-orbital segment. *Source:* From Ref. 23.

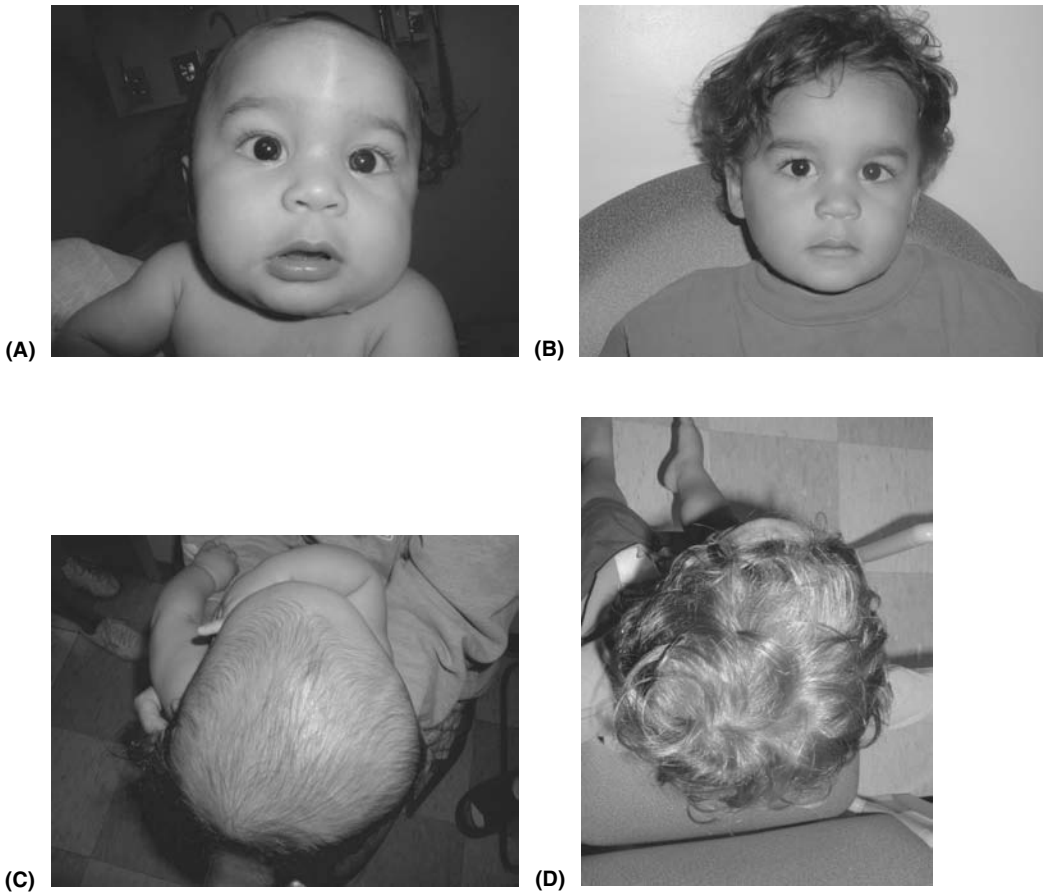


FIGURE 16 (A) Preoperative photograph of four-month-old male with metopic synostosis and trigonocephaly. (B) Postoperative photograph showing result two years after endoscopic correction. (C) Preoperative metopic synostosis with trigonocephaly. (D) Two years after endoscopic correction of metopic synostosis.

visualization. Having completed the lateral fronto-orbital and superior orbital osteotomies, a paramedian osteotomy is made along the metopic suture. The fronto-orbital segment can now be digitally outfractured and advanced (Fig. 20). Stabilization is achieved with a specially cut resorbable mesh as described for metopic synostosis (Fig. 21). The preoperative photographs of a six-month-old girl with right unicoronal synostosis are shown in Figure 22A, C, while the postoperative result at six months (Fig. 22B,D) and at one year (Fig. 22E,F) are displayed.

Representative pictures of actual patients are demonstrated. Figure 1 displays a minimally invasive sagittal suturectomy which will be followed by bifrontal and biparietal osteotomies. The *minimally invasive* planning/markings is demonstrated in Figure 16. Figure 17 shows an upper blepharoplasty incision being performed. The *minimally invasive* coronal osteotomy is demonstrated in Figure 21 while the fronto-orbital advancement is shown in Figure 18. After all the appropriate osteotomies, Figure 19 demonstrates an immediate intraoperative correction followed by resorbable plating shown in Figures 20 and 22. Figure 23 is a picture taken with the endoscope showing the completed osteotomy and means for dural inspection. We have increasingly turned to simple suturectomy (1 cm strip) for unicoronal synostosis and have found the results to be very favorable in mild to moderately severe cases.

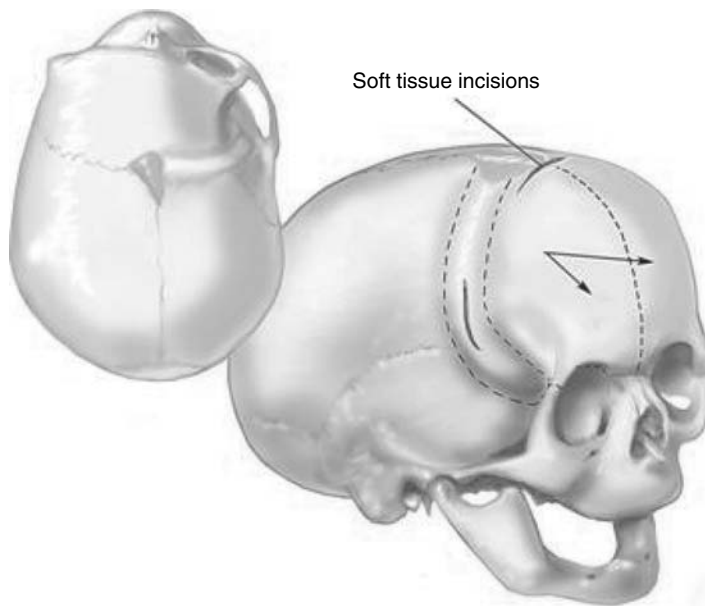


FIGURE 17 Artist's depiction of proposed incision sites and osteotomy cuts. With current practice, only one W-type incision is made lateral to the anterior fontanelle. *Source:* From Ref. 23.

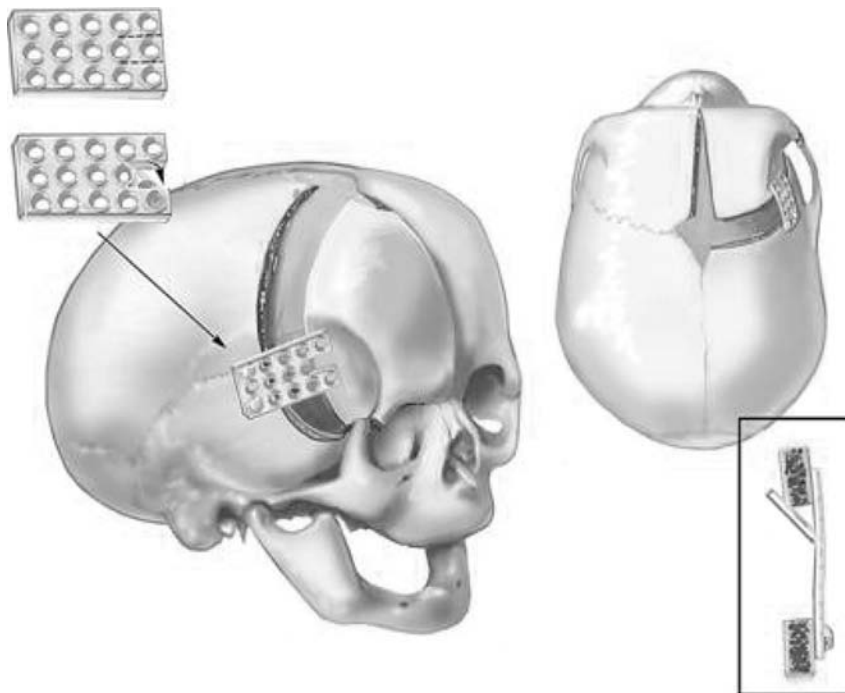


FIGURE 18 Blepheroplasty incision. *Source:* From Ref. 23.



FIGURE 19 Coronal suturectomy. *Source:* From Ref. 23.



FIGURE 20 Fronto-orbital advancement. *Source:* From Ref. 23.



FIGURE 21 Fronto-orbital advancement and fixation with specially designed, resorbable mesh. *Source:* From Ref. 23.

RESULTS

The Open Technique

Specific complications associated with craniosynostosis may be classified as either perioperative or postoperative. In a review of 204 patients treated for nonsyndromic craniosynostosis at Children's Healthcare Center of Atlanta, the overall perioperative complication rate was 9.8% (20/204).³⁴ Major complications were seen in total cranial vault remodeling for sagittal synostosis in which four experienced hypovolemic shock. One patient had a transection of the sagittal sinus, and one death was recorded. Seven patients were found to have syndrome of inappropriate antidiuretic hormone in the immediate postoperative period (four in the total cranial vault remodeling group). Other perioperative or immediate postoperative complications included severe chemosis (one case), wound infection (one case), and urethritis (one case). Perioperative complications, including air embolism after a venous tear, infarction, and damage to the unprotected brain, have also been reported (24).

Blood loss may be acute, but transfusions are usually necessary because of insidious losses throughout the case. Blood transfusions are required in almost all total cranial vault-remodeling procedures. Transfusion requirements have been reported from 15% to 90% of the patients' estimated red blood cell volume (25,26). Variability is seen based on the type of synostosis. Acute complications of blood transfusion are well known and include hypocalcemia, hyperkalemia, coagulopathies, and transfusion incompatibility. Delayed complications of blood transfusions usually involve the potential for transmission of viral infections.

A prospective statistical study of reoperation rates was reviewed in the treatment of 167 consecutive children with nonsyndromic and syndromic craniosynostosis over a 6-year period at Children's Healthcare of Atlanta (17). Mean length of follow-up was 2.8 years. Fronto-orbital remodeling with a floating forehead was completed at four to six months of age for nonsyndromic synostosis other than sagittal synostosis. This approach is similar to treatment of isolated synostosis in several centers (27–29). Bilateral fronto-orbital remodeling has been shown to be comparable with or better than unilateral remodeling and was done in all cases (28,30). Strip craniectomies were limited to sagittal synostosis with mild-to-moderate deformities. Total cranial vault remodeling was completed for severe deformities if the patient was

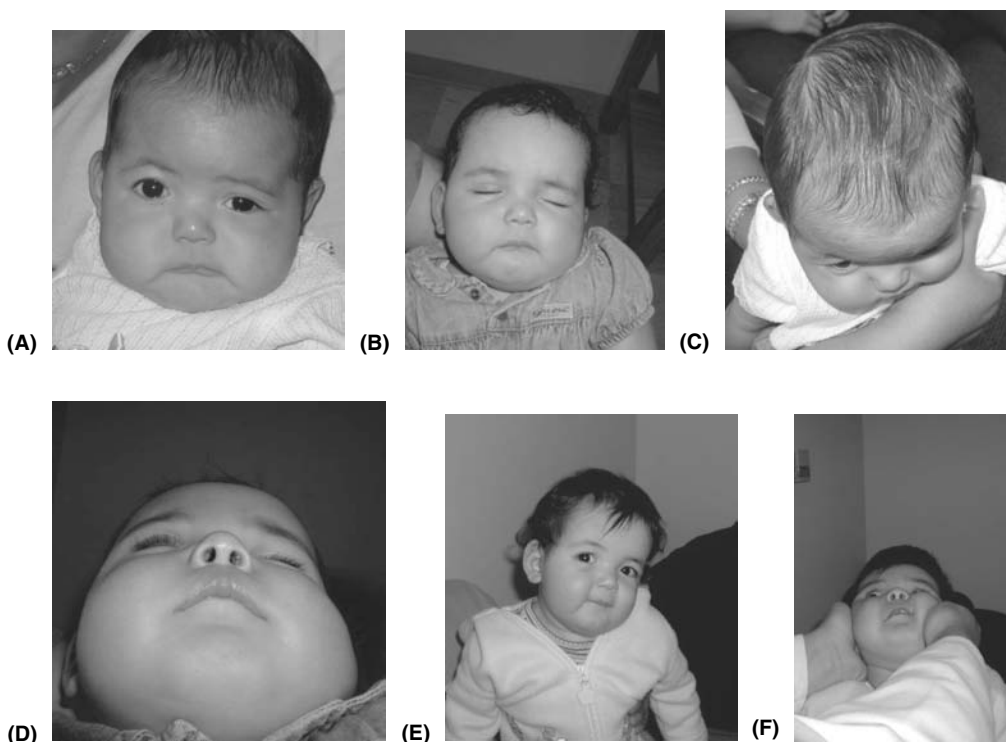


FIGURE 22 (A) Preoperative photograph of six-month-old girl with right unicoronal synostosis. (B) Postoperative result at six months. (C) Preoperative photograph showing right unicoronal synostosis. (D) Postoperative result at six months. (E) Postoperative result one year after endoscopic assisted correction. (F) Postoperative result one year after endoscopic assisted correction.

greater than seven weeks old. Patients with syndromic craniosynostosis underwent fronto-orbital advancement and cranial reshaping in four to six months unless increased cranial pressures required decompression.

Reoperation equal to or exceeding the magnitude of the original procedure occurred in 7% of cases. Total reoperation rates for syndromic and nonsyndromic synostoses were 27.3% and 5.9% respectively. Five of the 12 reoperative cases (41.6%) were completed for significant relapse, as demonstrated clinically and radiographically. Neither early nor late sagittal strip craniectomies required reoperations; only one patient in the late cranial vault-remodeling group for sagittal synostosis (>7 months) demonstrated relapse and required reoperation. Relapse was seen in two patients with bicoronal synostosis, requiring reoperation, and two more patients in this group underwent a second procedure for suboptimal cranial contouring.

Whitaker's classification (28) of clinical results after a craniofacial procedure includes Category III (C-III), requiring major bone grafting or other osteotomies, and Category IV (C-IV), requiring duplication of the previous craniofacial procedure. Total reoperations in our review were classified into the latter group. The increase in reoperation rates of syndromic was consistent with previous reviews. Whitaker (28,31) showed a C-IV reoperation rate of 3% for asymmetric lesions (isolated synostosis) and 64% for the symmetric lesions (95% in Apert syndrome). Excluding strip craniectomies, McCarthy had a 6.7% reoperation rate for isolated synostosis (16) and 28.3% for syndromic deformities (32). Surgical approaches similar to ours were used in each of the aforementioned studies.

In most studies, no differences were seen in reoperative rates for treatment of single-suture synostosis as related to age (23,34); recommendations for primary intervention ranged from 2 to 18 months. Wall et al. (27) showed an increased reoperation rate of 20% in



FIGURE 23 (A) Preoperative photograph of four-month-old girl with unicoronal synostosis. (B) Postoperative photograph six months after treatment with extended, endoscopic strip craniectomy. (C) Preoperative photograph of patient showing severe recession of the right supraorbital rim and frontal region. (D) Postoperative photograph showing marked improvement of fronto-orbital retrusion.

nonsyndromic synostosis when primary treatment was in patients less than six months of age compared with 5.6% in patients greater than six months old. In syndromic disorders, patients less than six months old had a 30.2% reoperation rate compared with 9.1% when older than six months (40.9-month follow-up). Other reviews have shown no relationship of age to reoperative rates (17).

The Endoscopic, Minimally Invasive Technique

The introduction of a minimally invasive approach for repairing craniosynostosis has decreased some of the risks and morbidity involved with the traditional open approach. When compared to a cohort of patients having open procedures at our institution over the same time period, our initial endoscopic assisted cases demonstrated a mean blood loss of 59 mL compared to 225 mL (73.7% decrease), which lowered the need for blood transfusions. When transfusions were needed during the endoscopic technique, about 40.6% less blood cells were used.

The mean length of operation using the endoscopic technique rather than the open technique also decreased from 156 to 83 minutes (46.8% decrease). Patients undergoing an

endoscopic operation recovered more rapidly and experienced a shorter hospital stay. The mean hospital stay decreased from 3.4 to 2.1 days (38.2% decrease).

Over the past five years, the endoscopic assisted, minimally invasive approach has been used. Seventy-six patients, ages 2.5 to 12 months have sufficient followup data to be included in this report. Of these 76 patients, 43 had sagittal synostosis, 21 had metopic synostosis, 8 had unicoronal synostosis, 1 had lamdoidal synostosis, and 3 had multiple suture synostoses. Operative times ranged from 25 to 184 minutes and were longer when the endoscopic assisted fronto-orbital advancements were carried out. Transfusions were needed in 61.1% of cases. No patient was admitted to the intensive care unit and all recovered on a monitored medical/surgical floor. Hospital stay ranged from one to four days, depending on whether the patient was from out of the region. Complications occurred in three patients. One had a dural tear which was easily repaired intraoperatively, one had erosion of the incision from the band, and one had bleeding in the recovery room with mild hypotension. The patient whose incision had eroded underwent successful debridement, while the patient with bleeding in the recovery room was transfused and had no further problems.

Follow-up times have ranged from one month to five years. Aesthetic results were satisfactory to excellent in all patients except for three. Reoperation was necessary in these three patients. One patient with unicoronal synostosis required open fronto-orbital advancement and two patients, one with lambdoid synostosis and one with multiple synostosis required an open posterior cranial vault remodeling.

DISCUSSION

As surgical techniques mature, emphasis shifts from postoperative survival to minimizing morbidity and minimizing surgical invasiveness. Traditional craniosynostosis repair has an established level of efficacy and safety, yet remains at most centers a major undertaking with significant blood loss, large scalp incisions, and significant postoperative swelling and duration of recovery. Increasingly, patients are becoming proactive consumers aware of surgical morbidities and options.

Recent reports have emphasized attempts to decrease morbidity such as techniques to limit blood loss. We feel the use of endoscopy allows our surgical team to minimize scalp incision, operative time, transfusion requirements, postoperative swelling and decrease hospitalization time. The degree of cranial reconstruction we can perform safely with direct endoscopic visualization and, in selective cases, absorbable fixation allows for a more extensive and immediate correction than suturectomy alone and minimizes postoperative banding. Many of our patients were placed in postoperative cranial bands/helmets to refine the initial result. Helmets and/or bands are well tolerated by both the infants and families and are used in many cases after endoscopic correction. It is important to point out that our patients requiring postoperative orthotics were in the helmet or band for relatively short intervals (mean 3.5 months) compared to some reports in the literature of band use up to 12 months and more following surgery (7).

We have seen no increase in intra- or postoperative morbidity with these techniques. Future refinements in our technique will hopefully allow us to translate a decrease in blood loss and transfusion amount to a decrease in absolute transfusion need. Our follow-up is relatively brief and will need to be expanded to further demonstrate technical long-term efficacy.

The preferred timing of surgery for the endoscopic approach is early, from two to four months. This lessens the complexity of surgery as the bones are more malleable and easy to cut and shape. Less surgery is necessary, using primarily simple strip craniectomies followed by helmet/band remodeling in the younger metopic, unicoronal and sagittal synostoses patients. In older children the bone is harder and more difficult to cut. There is also less head growth remaining, a prerequisite of the endoscopic, minimally invasive technique. Our practice has been to offer *minimally invasive* techniques to younger patients with mild-moderate deformity and *open* techniques for older and more severe deformities.

CONCLUSION

Over the past five years we have utilized modified *minimally invasive*, endoscopic techniques in the surgical correction of craniosynostosis. Added experience will further improve the operative times, blood loss, transfusion requirements, and complications. Furthermore, as specialized instruments are developed, these techniques will become easier and safer to perform. For selected patients, these techniques offer an alternative to traditional techniques in an effort to minimize postoperative morbidity and the need for prolonged cranial banding. Long-term follow-up will be needed to adequately assess the ultimate efficacy of these techniques. Traditional techniques for repair of craniosynostosis have historically an outstanding record of excellent aesthetic results with acceptable morbidity. Ultimately, each patient is best served with a customized plan developed and implemented by a multidisciplinary team capable of the full range of techniques.

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7 Syndromic Craniosynostosis

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INTRODUCTION

A *syndrome* is defined as a constellation of symptoms and signs that collectively characterize an abnormal condition or disease state, while *craniosynostosis* is defined as the premature fusion of the cranial sutures (1). This chapter will focus on the most common group of syndromic craniosynostoses and describe the salient points in their proper clinical and surgical management. As a group, the syndromic craniosynostoses may also be referred to as the craniofacial dysostosis syndromes. Both terms are often used interchangeably; however, the latter may better connote the scope of the deformity, which includes the cranial vault and the full extent of the facial skeleton.

The syndromic craniosynostoses do not fall easily into one category and make up a number of varied conditions resulting from multiple etiologies with a set of diverse factors that influence their pathogenesis. Earlier chapters have gone into greater detail as to the biologic principles of these disorders, however, a better understanding of the basis of each syndrome may have important implications in their ultimate clinical management. A sound insight into the variables that influence syndromic craniofacial malformations may well be a key predictor of growth abnormalities which could be invaluable in the development and application of surgical strategies tailored to each individual patient and to each individual diagnosis.

UNIQUE NATURE OF SYNDROMIC CRANIOSYNOSTOSES

Syndromic craniosynostoses result from an interaction between genetic factors, molecular and cellular events, mechanical and deformational forces, and secondary effects of each of these on normal growth and development (2–4). Despite the current explosion in the knowledge about the molecular biology of syndromic craniosynostoses, the same genotype has often been linked to multiple and diverse phenotypes with variable penetrance and expression (5,6). Patients carrying the same syndromic diagnoses often display remarkable differences in their level of severity and clinical outcome, perhaps confirming that the disease processes can occur through multiple pathways (2). The variability in both patient presentation and clinical outcome underscores the persistent complex interactions between underlying genetic and cellular predisposition and the local forces involved in growth and development.

Although many of these patients display a wide spectrum of phenotypes, the basic principles underlying the scope, timing, and approach to these complex craniofacial anomalies is based on the solid fundamental principles and tenants of craniofacial reconstruction. Patients with disparate syndromic craniosynostoses often undergo a similar pattern of reconstructive procedures in order to achieve improvements in the shape, symmetry, and balance of their face and cranial vault. The surgical approach to this challenging group of patients should be individualized with a surgical strategy formulated to address the overall craniofacial anomaly as well as the particular nuances in the deformation as manifested by each patient.

The surgeon often chooses from a stable of established operative approaches that can be modified as desired to suit particular patient needs. The timing of intervention of the staged procedures required for comprehensive craniofacial reconstruction of syndromic craniosynostoses should also take into consideration the individual needs of the patients; however, these immediate needs should always be tempered by the experience and knowledge of a trained craniofacial surgeon and pediatric neurosurgeon. Specifically, the impact of continued abnormal growth of the craniofacial skeleton cannot be underestimated as the surgical approach only deals with the structural aspect of the present abnormality and does not remedy the underlying growth disorder. The overall reconstructive goals of maximizing results, minimizing the number of operations, and achieving a predictable outcome should always guide the appropriate management for this complex group of patients.

SPECIFIC SYNDROMIC CRANIOSYNOSTOSES

Apert Syndrome

This syndrome, also known as acrocephalosyndactyly (Fig. 1), was first described in 1906 by a French neurologist and refers to patients who display characteristic features of craniosynostosis, midfacial hypoplasia, and symmetric compound complex syndactylysm of the hands and feet (7). The incidence of Apert syndrome is approximately 1 in 100,000 births (8). Although this occurs in a majority of patients sporadically there is an autosomal dominant inheritance pattern.

The synostotic presentation in these patients is usually simple and most often affects the coronal sutures bilaterally, resulting in a turribrachcephalic deformity and a short anterior cranial fossa (Fig. 2). The forehead is high, steep, and flat and is often associated with a transverse frontal skin furrow. The anterior fontanelle is unusually large with a lack of normal development of the adjacent metopic and sagittal sutures giving a characteristic bulging appearance, perhaps indicating a compensatory spatial shift of the brain (9). Other craniofacial features associated with this syndrome include hypertelorism, exorbitism, divergent strabismus, downslanting palpebral fissures, and ptosis with a characteristic "S" shaped deformity to the upper eyelid.

These patients usually have a high arched palate, a collapsed maxillary arch, and display an overt or submucous cleft palate in approximately 30% of the cases (10). There is an increased upper facial transverse width with acne vulgaris appearing in over 70% of

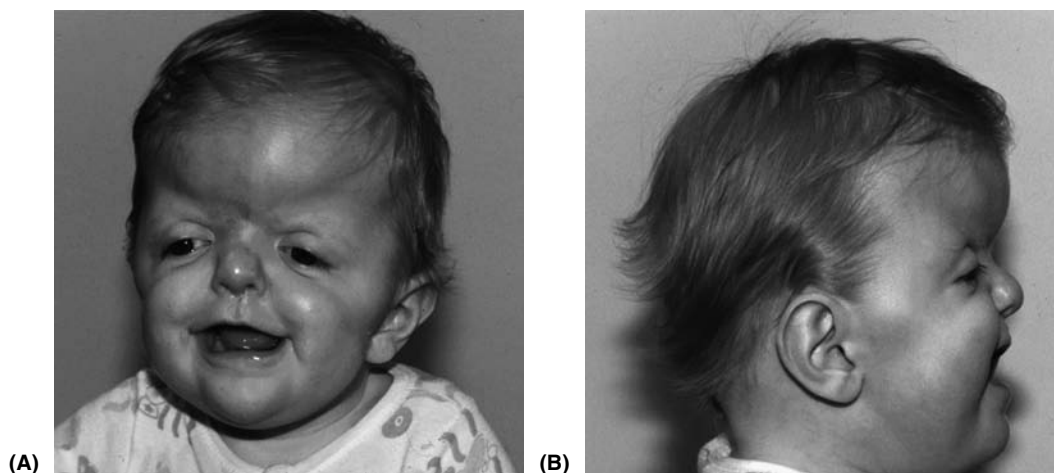


FIGURE 1 Clinical presentation of a child with Apert syndrome. (A) Frontal view and (B) lateral view.

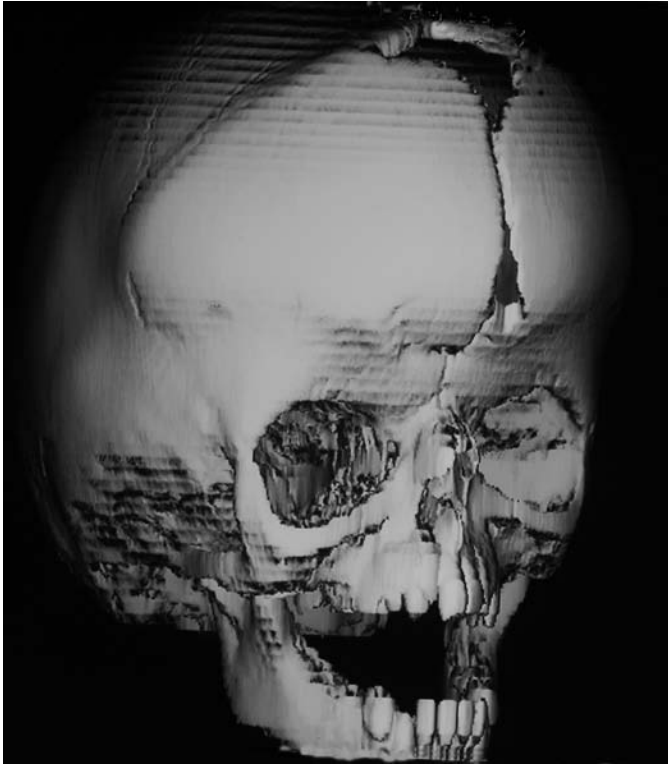


FIGURE 2 Three-dimensional computed tomography scan of a child with Apert syndrome. Note the synostotic coronal suture, the open fontanelle, and the midfacial hypoplasia.

patients in adolescence (11). Dental examination reveals a class III malocclusion with an anterior open bite and a pseudoprognathic mandible. In addition to the complex syndactyly, patients display radial deviation of the distal phalanx of the thumbs and a proximal delta phalanx (12). Apert syndrome is often associated with neuropsychological problems including attention deficit-hyperactivity disorder, developmental delays, and mental retardation (13).

Crouzon Syndrome

This syndrome was first described in 1912 by a French neurosurgeon who included four important physical findings of the disorder (14). These key findings included exorbitism, retromaxillism, inframaxillism, and paradoxical retrogenia. Patients with Crouzon syndrome also display characteristic features of craniosynostosis and midfacial hypoplasia (Fig. 3). The incidence of Crouzon syndrome is approximately 1 in 25,000 births (6).

Although a significant number of cases are thought to be sporadic, there is an autosomal dominant inheritance pattern with near complete penetrance and variable expressivity. The sutural synostotic pattern in these patients is usually simple and most commonly affects the coronal sutures bilaterally, resulting in a turribrachcephalic deformity with a short anterior cranial fossa (15). Crouzon's patients, however, have also displayed calvarial deformities such as scaphocephaly, trigonocephaly, and oxycephaly depending on the involved site of craniosynostosis. These patients can also show a predisposition to early or late craniosynostosis involving multiple sutures. Other craniofacial features associated with this syndrome include hypertelorism, exorbitism secondary to shallow orbits, and strabismus (16). These patients usually have a high, narrow, and constricted upper dental arch, crowded dentition; a class III malocclusion with a bilateral crossbite and an anterior open bite and



FIGURE 3 Clinical presentation of a child with Crouzon syndrome.

a pseudoprognathic mandible and retrogenia. There is a symmetric but variable degree of hypoplasia of the zygoma and maxilla. Crouzon syndrome is usually associated with normal intelligence.

Although there are some general similarities in the craniofacial features displayed by both individuals with Apert and Crouzon syndrome, close scrutiny of these patients will reveal distinct differences (17,18). The Apert patients often display a much more severe deformity, is highly associated with cleft palate, acne vulgaris, mental retardation, and the characteristic syndactylysm. Crouzon's patients usually display a less severe craniofacial dysmorphism, rarely have a cleft palate, do not display syndactyly, and often have normal intelligence.

Pfeiffer Syndrome

This syndrome was first described in 1964 and refers to patients who display craniosynostosis and the characteristic features of broad thumbs and broad great toes (Fig. 4), as well as occasional partial soft tissue syndactyly of the hands (19). There is an autosomal dominant inheritance pattern with complete penetrance and variable expression (20). The synostotic pattern in these patients is usually simple and most commonly affects the coronal sutures bilaterally, resulting in a brachycephalic deformity with a short anterior cranial fossa. Other craniofacial features associated with this syndrome include maxillary hypoplasia and a class III malocclusion. In addition these patients display hypertelorism, exorbitism, strabismus,



FIGURE 4 Characteristic features of the (A) broad thumb and (B) great toe of a child with Pfeiffer syndrome.

and downward slanting palpebral fissures (21). The degree of craniofacial deformity and the syndactyly associated with Pfeiffer syndrome is milder than the deformities associated with Apert syndrome, and the intellectual development is normal.

Carpenter Syndrome

This syndrome was first described in 1901 and refers to patients who display craniosynostosis and the characteristic features of polysyndactyly of the feet, brachydactyly of the fingers, clinodactyly, and occasional partial soft tissue syndactyly (20). Carpenter syndrome differs in the inheritance pattern from the previously described syndromic synostoses, in that the trait is autosomally recessive. The synostotic pattern in these patients most commonly affects the sagittal and lambdoidal sutures resulting in significant cranial asymmetry. Patients often display short stature and obesity. Mental retardation has been associated with this malformation (22).

Saethre–Chotzen Syndrome

This syndrome, first described in 1931, refers to patients who display craniosynostosis and the characteristic features of a low set frontal hairline, upper eyelid ptosis, facial asymmetry, and partial soft tissue syndactyly of the hands (20,23). There is an autosomal dominant inheritance pattern with high penetrance and variable expression. The synostotic pattern in these patients is usually simple and most commonly affects the coronal suture but may do so in an asymmetric fashion resulting in a plagiocephalic appearance (23). Intellectual development in these patients is usually normal.

Kleeblattschädel Anomaly

This calvarial deformity is made as a clinical diagnosis based on the phenotype (24). The skull takes on a trilobular or cloverleaf appearance (Fig. 5). It is a result of a complex, multiple cranial suture craniosynostosis. It may often be related to an underlying syndrome (frequently associated with Pfeiffer syndrome), but this is not exclusive and most often occurs sporadically. Other craniofacial features associated with this anomaly include severely recessed supraorbital rims, hypoplasia of the basal frontal bones, and marked convexity of the squamous portion of the temporal bones (Fig. 6) (20).



FIGURE 5 Clinical presentation of a child with a Kleeblattschädel deformity. (A) Preoperative frontal view. Note the trilobar appearance of the skull. (B) Postoperative frontal view, (C) preoperative cranial view, and (D) postoperative cranial view.

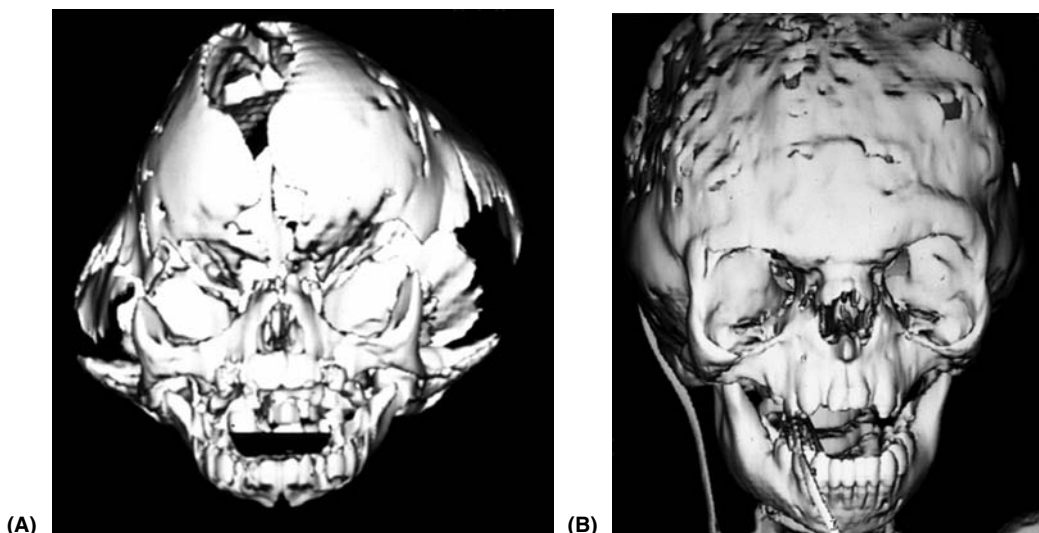


FIGURE 6 Three-dimensional computed tomography scan of a child with Kleeblattschädel deformity. (A) Preoperative view and (B) postoperative view.

DIAGNOSIS AND INITIAL CLINICAL EVALUATION

When first confronted with a patient that may be afflicted with a syndromic craniosynostosis, an organized and thorough approach is mandatory. A comprehensive and systematic evaluation is necessary in order to arrive at the appropriate diagnosis, fully assess any associated medical concerns, and embark on the proper clinical management of the patient. A careful functional and aesthetic assessment is vital to a successful outcome and is best ascertained through a focused but detailed history and physical examination.

Both a trained craniofacial surgeon and an experienced pediatric neurosurgeon best address the initial evaluation of these difficult patients. The expertise of these two accomplished subspecialties complement each other by combining their individual talents and skills to treat the multifaceted concerns of children afflicted with these complex deformities. The pediatric neurosurgeon is charged with discerning the best approach to care for the impact of the structural anomalies, intrinsic to the syndromic patient, on the central nervous system. Associated issues such as hydrocephalus and Chiari I malformation must be recognized. It is extremely important that elevations in intracranial pressure (ICP) be recognized early. Such assessment of ICP is best evaluated by a pediatric neurosurgeon. In addition, the imaginative craniofacial surgeon will rely on the proficiency of their pediatric neurosurgical colleague to confine them to the realm of the possible, emphasizing safety and tempering creativity by stressing practicality and discerning the limitations of the brain.

Although the anatomic features of the craniofacial anomaly may often be immediately obvious in children with syndromic craniosynostosis, the detection of significant associated problems impacting each individual patient may be much more subtle. A targeted but thorough history is imperative in order to focus the physical examination, formulate a more specific diagnosis, and outline the correct clinical course of management. A family history or a family resemblance of the child to a close or distant relative connotes an inherited trait and could be invaluable in making a more specific diagnosis as well as place certain physical attributes into better perspective. A birth history can give clues as to possible complications around the time of delivery such as episodes of anoxia, infections, or trauma. Exposure to drugs, radiation, alcohol, or other harmful substances needs to be documented and may be an important factor in patient management. A history of headaches, lethargy,

vomiting, or difficulty seeing or following objects could be clues as to underlying neurological disorders. In addition, seizure activity and developmental delay indicate abnormal brain function.

Difficulty feeding may signify a problem with maternal bonding or may be an understated sign of respiratory distress. Snoring, noisy breathing, or significantly worsening airway obstruction with colds could indicate an underlying problem with sleep apnea. The midfacial hypoplasia in these patients can be quite severe and can often lead to a spurious diagnosis of choanal atresia by the neonatal intensivist after difficulty placing a nasogastric tube.

The physical examination of the syndromic craniosynostotic patient is of particular importance as it helps to establish the diagnosis, identify the extent of the deformity, and form the basis of a treatment plan and surgical strategy for reconstruction. A comfortable familiarity of the associated physical findings that distinguish the common craniofacial syndromes should guide a vigilant examination of the patient. An initial skilled general survey of a child with syndromic craniosynostosis should detect and confirm readily apparent physical findings, while proficient powers of observation are needed to appreciate the subtler characteristics of abnormal facial form inherent in a comprehensive analysis of craniofacial dysmorphology. Attention to detail will be rewarded with a clearer picture of the patient's current status, an outline of their structural and functional deficits, and a working classification of the deformity. The examiner should be alert to any growth disturbances, neurologic deficits, and developmental delays displayed by the patient, and assess basic functions such as the child's ability to see, eat, and breathe. Basic problems in any of these areas will significantly impact the appropriate medical and surgical management of the patient and will guide appropriate consultation and further diagnostic workup.

A head circumference should be taken on all children in their evaluation for a syndromic craniosynostosis. A small head could connote a primary growth abnormality of the brain or an uncompensated growth restriction of the skull secondary to craniosynostosis. A large head could indicate hydrocephalus or a compensated growth pattern of the skull secondary to craniosynostosis. Palpation of the cranium should include the fontanelles and the cranial sutures. An enlarged fontanelle is commonly seen in Apert syndrome, while a bulging fontanelle could indicate increased ICP and possible hydrocephalus. A palpable ridge can suggest synostosed sutures as could premature closure of the fontanelles.

The general shape of the head is an indication of the underlying pattern of craniosynostosis and is perhaps the most important factor in establishing a clinical diagnosis. Brachycephaly (short and flat head) and turriccephaly (tall head) results from bilateral coronal craniosynostosis and are the most common findings in Apert's (Figs. 1 and 2), Crouzon's (Fig. 3), and Pfeiffer syndrome. Kleeblattschädel presents as a trilobed skull and indicates multiple suture synostosis (Figs. 5 and 6). Trigonocephaly (triangular head), scaphocephaly (keel shaped head), and plagiocephaly (slant head) can occur but are less common in syndromic cases. A low set hairline is a component of Saethre-Chotzen syndrome. A close analysis of the periorbital region and the eyes is a vital portion of the physical examination. Inspection of the globes will often reveal exorbitism secondary to hypoplastic orbits and in extreme cases may result in corneal exposure posing a risk to eyesight (Fig. 3). Hypertelorism and telecanthus are prominent findings in the craniofacial dysostosis syndromes and can be more accurately ascertained by a measurement of intercanthal distance. Intraocular assessment of the fundus and visualization of the optic disk should be sought to rule out pallor and papilledema; signs of increased ICP. Congenital ptosis is also an element in the diagnosis of Saethre-Chotzen syndrome.

Malar flatness and midfacial retrusion is a common constituent in syndromic craniosynostosis and may be secondary to growth disturbances involving the cranial base. The midfacial contraction and concave profile can be severe in these patients and can result in respiratory distress as the posterior pharyngeal wall comes in close proximity to the posterior palate obstructing the normal free flow of air through the nasopharynx. Malar width is characteristically increased, while sagittal projection in the midline is decreased leading to a wide, dishface appearance. Intra-oral examination may reveal a cleft palate or more commonly a high arched palate. These patients often display a class III malocclusion with an anterior open bite and a posterior-lateral crossbite. Both the occlusal and palatal malformations can result in

abnormal speech. Evidence of persistent torticollis could signal cervical spine deformities, an extra-ocular muscle problem, structural foreshortening of the sternocleidomastoid muscle, or simply a habitual inclination. The recognition, diagnosis, and treatment of this problem are of paramount importance as the muscle imbalance can result in significant asymmetry affecting the growth and development of the face and cranium.

An evaluation of the extremities is essential to the diagnosis and comprehensive treatment of the children with syndromic craniosynostosis. Syndactyly is a prime component of Apert syndrome while abnormalities of the thumbs and toes are elements of Pfeiffer and Carpenter syndrome (Fig. 4).

TESTS, CONSULTATIONS, AND CLINICAL MANAGEMENT

Imaging studies are an invaluable aid in the evaluation of the patient with syndromic craniosynostosis. A skull series is helpful to scrutinize the cranial sutures and detect radiographic evidence of fusion. Computed tomography (CT) is an essential study to assess the structural anatomy of the brain and surrounding cranial vault. A thorough evaluation of the scan is performed looking for evidence of hydrocephalus, signs of increased ICP, and indications of pathologic neuroanatomy. The scans also provide an exceptional examination of the bony anatomy of the craniofacial skeleton. Three-dimensional reformatting may convey a clearer picture of these complex deformities and can also serve as an effective teaching tool (Figs. 2 and 6). Magnetic resonance imaging can be complementary to a CT scan by furnishing a distinct view of the brain and is particularly helpful in assessment of Chiari malformations.

A neuro-ophthalmologic consultation for a careful dilated examination of the eyegrounds can be an important adjuvant to the initial cursory view of the fundus under suboptimal conditions. A dilated exam will document the baseline status of the optic disc and may impact the timing of surgical intervention. A genetics consultation will refine, confirm, or help establish the diagnosis as well as screen for associated problems and suggest further workup or necessary testing. Genetic counseling can console and inform the patient's family and generate a letter as a reference to document the inheritance patterns and subsequent risk to future offspring for both the parent and the child. Rapid progress in the discovery of associated mutations for many of the craniofacial syndromes has resulted in blood tests that have been developed to assist the geneticists in arriving at more accurate and specific diagnostic classifications.

When there is a question as to breathing difficulties, a sleep study can be quite helpful to rule out central or obstructive sleep apnea. Pulmonary consultation may be valuable to assist in judicious conservative medical management in an attempt to avoid or postpone an operation or as a provisional step prior to surgical intervention. Difficulties with feedings may also be due to underlying breathing problems, or alternatively, may be due to a variety of other etiologies such as bonding issues, trouble swallowing, or just impediments from technique or equipment. A feeding specialist can provide support and assist parents in correcting technique, recommending changes in equipment, or helping to determine more functional causes for difficulty feeding.

Neuropsychological evaluation is useful in determining baseline function and in detecting cognitive deficits. The evaluation can serve as a comparison to the patterns of future development or, more specifically, can lead to recommendations for intervention programs early on. Neurologic disorders, such as seizures, are best managed with the assistance of a neurologist while physical developmental delays may benefit from consultations with rehabilitative medicine and require the early initiation of physical therapy. Children with syndromic craniosynostoses often have multiple related medical problems and are particularly well suited to a multidisciplinary team approach to their care, frequently requiring close interactions between medical and dental specialists as well as those in the allied health professions. A social worker is an important element of the team and can advise the parents in regards to issues of insurance coverage through private and governmental programs. The social worker can also assess the family's ability to cope and assist in creating a support system capable of dealing with the deluge of social and medical responsibilities incumbent in raising a child with a syndromic dysostosis. A speech pathologist is an integral part of the team, and can monitor speech development and act as an advocate for the intervention of school and community-based

programs as well as monitor the quality of help delivered by those programs. An audiologist can monitor hearing and be a liaison to the ENT specialists, following the progress and success of myringotomy tube placement, and highlighting the need for early screening such as audio evoked potentials.

The dental specialties play an integral role in the craniofacial team and work in concert with the craniofacial surgeon to exact a superior level of comprehensive rehabilitation. Pediatric dentistry is an essential component in the preventative care of these patients as the children often exhibit early malocclusion, poor hygiene, and dental caries. Preemptive care is a key to maintaining the fitness of deciduous and newly forming permanent dentition allowing the successful bio restoration of oral health. The orthodontist plays a fundamental and crucial role in combining facial analysis with an assessment of tooth malposition to arrive at a treatment plan that complements and supports the surgeon in achieving a superior craniofacial reconstruction. Close interaction between the surgeon and the orthodontist throughout the growth and development of the syndromic patient will promote cooperative strategic interventions by both specialties. Orthodontic phased treatment plans may include palatal expansion, attention to crowding, tooth extraction, and placement of a retainer.

EVOLUTION OF SURGICAL MANAGEMENT

The surgeon's goals are to provide a predictable and durable reconstruction that restores facial form, reestablishes symmetry, and attains the best results in the least number of operations. All of these objectives must be considered with regard to the safety of the patient and the inconsistent and irregular nature of growth in the child with syndromic craniosynostosis. Despite the array of surgical procedures developed to reconstruct these children, the deleterious effects of abnormal growth persist, suggesting that operative intervention is more effective at treating the symptoms of the underlying disorder rather than the disease itself. Growth, therefore, acts more as a nemesis than as an ally in the struggle to achieve a long lasting reconstruction. A successful surgical strategy must consider the tactical points in time during a child's growth and development that operative intervention will best realize the ultimate goals while still addressing the social concerns of each patient. The challenges of continued anomalous growth must be balanced by the psychosocial concerns of the child affected by syndromic craniosynostoses. These children still suffer from severe disfigurement during very impressionable stages of development that are critical in shaping self esteem, influencing self confidence, and impacting social competence.

Surgical intervention must also address any pressing health concerns such as high ICP, airway obstruction, or severe exposure of the globes refractory to medical management. These health concerns will supplant any treatment protocol or timing philosophy for operative intervention in the child with syndromic craniosynostosis. Hydrocephalus can develop at any point along the course of treatment and must be recognized and treated quickly. There is a higher incidence of increased ICP in children with multiple suture craniosynostosis and an increased number of involved sutures in the syndromic patient with craniosynostosis (25) particularly in patients with cloverleaf deformities. Therefore, it is imperative to be vigilant and maintain a high index of suspicion for any sign or symptom of high ICP. In cases where there may be a question of the status of ICP, a monitor can be placed surgically and the patient evaluated for a few days in an ICU setting to get an idea of the range of pressure throughout the day and in relation to particular activities and times. High ICP will usually mandate acute surgical intervention targeted at the etiology; this could mean the placement of a shunt, the expansion of the cranial vault, or a combination of both. Close communication between the pediatric neurosurgeon and the craniofacial surgeon is essential in cases of high ICP so that an appropriate operation is performed and so that the timing of the operation is optimal.

Airway obstruction requires the interaction between the craniofacial surgeon, the ENT surgeon, and the pulmonologist. An acute structural problem can require an emergent tracheostomy, while less severe airway compromise may be amenable to continuous positive airway pressure therapy, or supplemental oxygen and close monitoring. There is a high incidence of airway difficulties in the syndromic patient with craniosynostosis because of the more global effects of growth restriction on the midface and the increased incidence of

associated malformations found in children with these disorders. Therefore, it is imperative to be vigilant and maintain a high index of suspicion for any sign or symptom of airway obstruction or sleep apnea. A subacute or slowly worsening picture of airway obstruction may be best treated by surgical intervention aimed at the inciting structural impediment. In the case of a severely retruded midface, a bony advancement could significantly open up the area between the choana and the posterior pharynx and have a salutary effect on the airway. Mandibular advancement or advancement of the chin with or without a hyoid suspension could move the tongue forward and provide more room in the posterior airway. Lesser procedures targeted at specific problems such as septal deviations, excess local tissue in the palate or tonsillar area, or other extrinsic or intrinsic obstructions should also be considered as appropriate after a detailed workup. All of the options for airway difficulties should be considered in terms of the acuity of the problem and in terms of the timing of the patients staged reconstruction in order to minimize operations and maximize results. A particular operation may fit well into the overall scheme of reconstructive options for a specific patient but airway concerns may require that the timetable be moved up so as to accomplish two goals at once improving both the structural and function outcome of reconstruction.

Exposure of the globes requires close interaction between the craniofacial surgeon and the ophthalmologist. Corneal desiccation and injury can result from the inability of the eyelids to adequately cover and protect the globe. The problem becomes more pronounced at night and can lead to a severe diminution in visual acuity over time. Treatment may include patching, topical applications, and tarsorrhaphy. In some severe cases, the diminutive orbit cannot accommodate the growing eye and periorbital contents leading to herniation and even dislocation from the orbit. There is a higher incidence of globe exposure in the syndromic patient with craniosynostosis because of the more global effects of growth restriction on the midface and as well as the recession of the frontal-orbital bar. A subacute or slowly worsening picture of globe exposure may be best treated by surgical intervention aimed at increasing orbital volume. In the case of a severely retruded frontal-orbital bar or midface, a bony advancement could significantly expand orbital size and capacity allowing the globes and periorbital contents to settle back into the confines of the bony socket (Fig. 7). Procedures that expand the lateral orbital wall or "blow out" the orbital floor can also achieve similar success in less severe cases. Close ophthalmologic monitoring is imperative to assure appropriate management. Again, all of the options for dealing with globe exposure should be considered in terms of the acuity of the problem and in terms of the timing of the patients staged reconstruction in order to minimize operations and maximize results.

The timing of the release of the cranial sutures in the patient with syndromic craniosynostosis has been controversial. The benefits of the early release of the cranial sutures have more to do with the philosophy of the surgeons as to the impact of craniosynostosis on the brain and the benefits and disadvantages of a more delayed approach to the durability of the overall reconstruction. There is no doubt that a documented problem with an increased ICP would mandate intervention; however, short of such an indication, there is disagreement as to the surgical timing of the initial attempt at reconstruction in the syndromic patient.

The history of early reconstruction (neonatal–three months) was predicated on a belief that the synostotic suture was a principal participant in the expression of the abnormal phenotype. It was thought that by releasing the synostotic suture the calvarial vault and the frontal-orbital bar could now take on a normal growth pattern. The expanding brain, unfettered by the growth restriction derived from the fused suture, would thrust the facial skeleton forward, directing and guiding future development, and inducing a normal growth pattern. This philosophy proved untrue as a simple suture release was remarkably unsuccessful in preventing the significant sequelae and seeming inevitable disfigurement associated with the growth and development of the craniofacial skeleton in these syndromic patients. The surgical philosophy then changed to a more comprehensive strategy aimed at releasing the synostotic sutures while concomitantly relocating the bones of the cranial vault into a more anatomically correct position. This strategy, again, called for performing the operation very early on, allowing the brain to expand and impel the well reconstructed cranial vault and frontal-orbital bar forward, inducing a normal growth pattern. Even more aggressive surgical teams attempted early monobloc procedures that combined the reconstruction and advancement of the cranial



FIGURE 7 A child with Crouzon syndrome displays exorbitism, hypertelorism, and midfacial retrusion, and undergoes a reconstruction using the monobloc procedure. (A) Preoperative frontal view, (B) postoperative frontal view, (C) preoperative lateral view, and (D) postoperative lateral view.

vault, the frontal-orbital bar, and the midface in one piece at a very early age. These procedures also “took advantage” of the propensity for the expanding brain to promote normal growth and development of the craniofacial skeleton. The unacceptable levels of both morbidity and morality that were associated with its application in the infant marred any success of the cases using the monobloc technique. The literature seems to document, however, that even the more conservative operations had a significantly higher incidence of revisional surgery and poor results in the syndromic patient when compared to the nonsyndromic patient (15,26,27). The higher incidence of revisional surgery is an important outcome measure as the number of operations should be minimized not only to subject the patient to less risk but also because the quality of the bone decreases each time it is subjected to a surgical procedure. These findings again suggest that the deleterious effects of abnormal growth endure, and as stated previously, that the operative intervention is more effective at treating the symptoms of the underlying disorder rather than the disease itself. The premise that the release of the brain will allow a normal pattern of expansion and growth is most probably a faulty one. The entire cranial vault and facial skeleton seem to be affected by the syndromic craniosynostoses and may have much of their pathologic growth disturbances as a manifestation of the nearly inaccessible sutures at the cranial base. Early release of the accessible craniosynostotic sutures, therefore, without a specific medical indication, as outlined previously, seems doomed to failure. On the other hand, waiting too long to initiate reconstruction of the cranial vault and frontal-orbital bar could also have a deleterious effect on the eventual outcome of a phased reconstructive strategy. Allowing the abnormal growth pattern of syndromic craniosynostosis to go unchecked could result in a craniofacial skeleton that is so skewed and misshapen as to make reconstruction even more difficult to get back to baseline. Furthermore, the transient osteogenic capability of the dura to heal bony defects should be taken advantage of, as it precludes the need for extensive bone grafting. These bone grafts will most definitely be needed down the road for future reconstruction and should be conserved whenever possible. Finally, the initial craniofacial reconstruction is also technically easier at a younger age, as the bones are more easily cut and shaped allowing a more accurate anatomical restoration. With adequate expansion, brain compression can be avoided and particularly with adequate posterior reconstruction, there can be improvement in cerebellar ectopia or Chiari malformation. The surgical philosophy driving the timing of the release of the cranial sutures in the patient with syndromic craniosynostosis, therefore, needs to balance the desire to attain a normalization of morphology with the realization that the propensity for abnormal growth still persists. The compromises needed to achieve that balance can best be made on an individual basis with careful consideration of any pressing health concerns, such as high ICP, which would necessarily direct surgical intervention.

CURRENT SURGICAL THERAPY AND TIMING

The correction of the deformities of a child with syndromic craniosynostosis is usually accomplished through a series of operations separated over time with respect to the growth and development of the craniofacial skeleton. This phased treatment plan is based on the differential growth patterns of the cranial vault, the midface, and the mandible. The first operation done for the syndromic patient with craniosynostosis is usually a frontal-orbital advancement. Depending on the degree of the deformity and the syndrome involved, some sort of cranial vault reshaping is usually performed simultaneously (Fig. 8). A child with Saethre-Chotzen syndrome may manifest a purely cranial dysmorphology with respect to the bones of the craniofacial region. In this case, the surgical intervention would be limited to the affected area and there may be no need for further operations. Alternatively, a child born with severe multiple suture synostosis, manifesting itself as a Kleeblattschädel anomaly, will often require an emergent surgical intervention aimed at reducing the increased ICP, expanding the cranial vault, and reshaping and advancing the frontal bone and supra-orbital bar (Figs. 5 and 6). Such a case is usually just a first phase of a presumed staged reconstruction to be carried out throughout the course of the child's growth and development. Similarly, children with Apert, Crouzon, and Pfeiffer syndromes will most often require a staged approach to reconstruction. The timing of the frontal-orbital advancement in these patients is usually delayed, in our institution, until

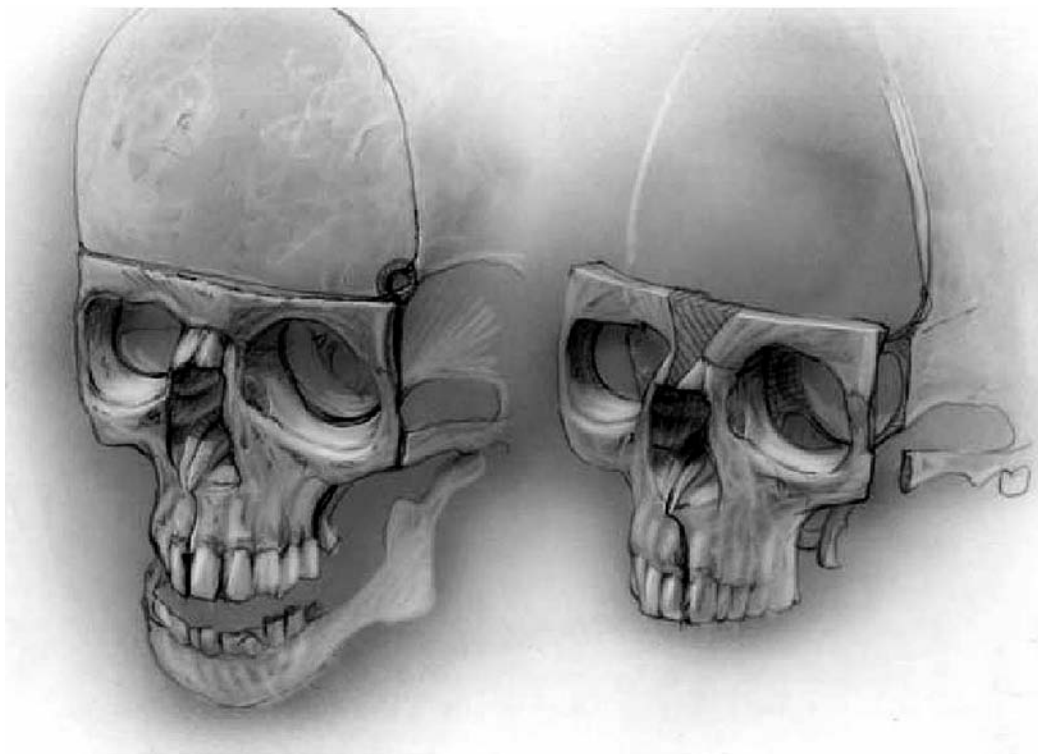


FIGURE 8 A schematic depiction of a frontal-orbital advancement and cranial vault reshaping procedure. The technique for addressing a turriccephaly deformity is presented.

9 to 12 months of age to achieve a more durable reconstruction and to minimize revisional surgery as outlined in the last section. The timing of surgery is clearly decided on a individual basis and is strongly influenced by concerns regarding intracranial crowding.

In the case of a child with significant turriccephaly, most often seen in children with Apert syndrome as well as some with Pfeiffer and Crouzon syndromes, the paradigm of reconstruction may need to be altered. In a turriccephalic deformity there is a shortening of the cranial base and an increase in the vertical height of the forehead and the cranial vault. The correction of the tall forehead and cranial vault will require additional room to allow the brain to sit comfortably within the confines of a more spherical cranium. The additional room is best attained through a posterior cranial vault expansion (Fig. 8). The expanded posterior cranial vault combined with a frontal-orbital advancement should allow adequate room for the displacement of the heightened skull and underlying brain to settle into after reconstruction. The posterior cranial vault expansion can be performed simultaneously if the patient is placed in the "sphinx" or "seal" position on the operating room table. Preoperative assessment of the neck as well as ruling out a Chiari malformation is mandatory if such a position is to be used since the neck is placed in extreme extension during positioning. Alternatively, the two operations can be staged, allowing an aggressive posterior cranial vault expansion in a prone position at six to nine months of age followed in three to six months by a frontal-orbital advancement. This approach is modified as dictated by any concerns for increased ICP. Dealing with a shunt during a craniofacial procedure requires careful dissection. If possible, the shunt is best placed posteriorly to allow easy access to the frontal portion of the head. If reconstruction is necessary around the shunt, we prefer to keep a layer of galea and soft tissue over the shunt, thereby maintaining a biological barrier. If reconstruction of skull around or near the shunt is necessary, a small island of bone can be kept under the shunt and later secured to the reconstructed skull to stabilize the shunt.

Frontal-Orbital Advancement and Cranial Vault Reshaping

The patient undergoes a similar preoperative assessment as has been outlined previously in other chapters for an intracranial procedure. The child is placed in the supine position with the endotracheal tube centered out of the mouth. The operative table is set at 90 to 180 degrees away from the anesthesiologist. A serpentine bicoronal incision is made from the back of the ear on one side to the other (Fig. 8). A suprapariosteal dissection is performed until approximately 1 cm above the superior orbital rims. The dissection is then carried subperiosteally to the orbits and the periorbital is degloved from the globes medially just above the canthus and laterally just below the zygomaticofrontal (ZF) suture. Depending on the degree of the deformity, the temporalis muscle is taken down to expose the temporal fossa and allow repositioning of the muscle. An arching incision with a cautery is made in the periosteum well above and behind the temporal line. A subperiosteal dissection is made rotating the temporalis anteriorly and medially.

A back cut of the muscle at the level of the temporal portion of the zygoma may be necessary to gain the mobilization for medial advancement. A bi-frontal craniotomy is performed by the neurosurgeon, with care being taken to avoid damage to the dura and particularly the underlying sagittal sinus. The bifrontal craniotomy should extend down to within a centimeter of the orbit. If the craniotomy extends down directly to the orbital roof there may be too small an orbital bar left. If the craniotomy is too high, there can be too much residual bone to be able to safely elevate the frontal lobes extradurally, without excessive retraction and therefore damage. Mannitol (1 gm/kg) is generally given at the beginning of the craniotomy and pCO₂ is carefully monitored. Each of these maneuvers relaxes the brain and makes retraction more gentle. Careful extradural dissection of the subfrontal region allows for safe creation of the orbital bar. Dural tears can occur during craniofacial procedures and usually require just a primary closure of the tear. Tears in the sagittal sinus can be life threatening, particularly in young children, and require aggressive management. Care must be taken during reconstruction to avoid kinking of the sinus. Sagittal sinus thrombosis must be avoided as attempts are made to alter the overall calvarial shape.

An osteotomy is then performed at a level just below the ZF and carried along the orbital roof to the midline; this is met by a similar osteotomy coming from the opposite side. The supra-orbital bar is then removed. The osteotomy is performed while protecting the globes and the brain with malleable retractors. The cranial bone from the craniotomy is taken to a back table and split with an osteotome if possible to secure additional bone for grafting. The temporal fossa is then approached and a barrel stave osteotomy is performed. Moving posteriorly, barrel stave osteotomies are also performed along the parietal skull depending on the degree of deformity of the cranial vault. The barrel stave osteotomies are then out fractured and molded with a bone bending forceps. Tactical cuts are made in the out fractured bone to allow the parietal and temporal region to take on a more normal anatomic appearance. If there is a significant element of proptosis, the periorbital can be incised allowing some of the fat to herniate. The frontal-orbital bar is then bent into an anatomic shape and then placed in an advanced and lowered position, and fixed at the lateral orbital rim with plates and screws. An advancement of at least 1 to 1.5 cm preferred as overcorrection is often necessary to counter the recoil forces of the surrounding soft tissue and the predisposition for recession of the orbital bar over time. The soft tissue in the coronal flap can be scored and released to lessen the resistance to advancement. Metal microscrews are used or alternatively resorbable systems can be substituted, but the advancement calls for structural integrity, so strength of the fixation is paramount. An interpositional bone graft is then placed to fill the gap laterally. If a thin piece of bone graft can be split, it can be used as a plate and fixed to the lateral orbital rim and the lateral portion of the supraorbital bar. It is important to have a posterior sloping trajectory of the orbital rim to keep the incline at the forehead more acute; this should help to reduce the propensity for the forehead to become turricephalic over time.

The bones of the forehead are then cut, bent, and formed into an anatomic shape. The bones are then fixed to the supra-orbital bar. A more acute angle of the forehead is sought, as is a slightly exaggerated transverse curve as an overcorrection against the susceptibility to developing a brachyturricephaly over time. A gap of at least 2 cm is maintained between the

posterior portion of the frontal bone and the anterior border of the remaining margin of the craniotomy, so as to prevent a recurrent synostosis.

The decision to perform a lateral canthopexy varies from surgeon to surgeon. Some perform a lateral canthopexy in all frontal-orbital advancements, others are more selective. At our institution we determine the need for a canthopexy clinically. Usually, if the lateral canthus looks good, preoperatively it will reattach in a normal position; however, if there is a poor position of the lateral canthus preoperatively or an intraoperative question arises as to malposition, the canthopexy is performed. The technique uses a 3.0 clear nylon suture and requires the careful isolation of the lateral canthal mass by grasping the lateral canthus externally with a forceps. The needle is then driven through the skin, from inside to out, through the area that was being grasped. The needle is then introduced back through the same hole at a different trajectory and pulled back and forth to fix the lateral canthal mass but release any skin attachment to the suture. The suture is then pulled and attachment to the lateral canthus is checked, the lower lid should move freely when the suture is pulled. The suture is then fixed behind the advanced lateral orbital rim, or through a separate set of drill holes, pulling the canthus laterally and posteriorly. Attention is then directed to the temporal fossa, which is shaped with a combination of bony remolding and muscular reposition. The temporalis is fixed with 3.0 absorbable suture. The frontalis is scored to release any "memory" of the abnormal forehead and allow a more natural drape of the soft tissue. The coronal flap is securely closed with interrupted absorbable galeal sutures and the scalp is closed with closely placed 3.0 plain running sutures. A headwrap is applied and every attempt is made to extubate the child in the operating room.

Postoperatively, the patient is sent to the intensive care unit where vital signs and urine output are monitored closely. We prefer to use normal saline as the maintenance intravenous fluid to help guard against hyponatremia. Sodium levels and hematocrit are followed on a regular basis. In challenging cases where extensive dissection around the brain is required such as a Kleeblattschädel anomaly, phenobarbital may be used to prevent the possibility of seizures. Narcotics are used sparingly and judiciously so that an accurate neurologic examine can be followed. The child is usually transferred to the floor on the second postoperative day and is discharged in five days after the extensive periorbital swelling has decreased.

Turriccephaly

As stated previously, some children with Apert syndrome and, less often, children with Pfeiffers or Crouzon syndrome, will exhibit severe turriccephaly. In those cases, a simple frontal-orbital advancement will often not be sufficient to address the extreme height of the cranium and forehead. Comprehensive reconstruction requires shortening of the cranial vault and forehead, necessitating an expansion of the length and width of the skull in order provide enough room for the brain as it displaces into a more normal anatomic position.

The operation for turriccephaly will include both a frontal-orbital advancement and a posterior cranial vault expansion combined with a shortening of the height of the cranial vault (Fig. 8). The procedures can be combined by using a "sphinx" or "seal" position or staged as outlined previously. If staged, we perform the frontal portion of the operation first and extend as far posteriorly as can be done safely. The need for a second stage is then carefully monitored clinically based on the result and subsequent craniofacial growth. The posterior cranial vault expansion requires a craniotomy for access to the posterior cranial vault. A portion of the superior cranial vault can be left in place along the midline as long as there is adequate access to the parietal and occipital bones or alternatively, the bone from the craniotomy is removed on bloc allowing generous exposure to the posterior and lateral cranial vault. With careful attention to the region of the torcula and the sigmoid sinus, the brain is retracted with a malleable retractor and barrel stave osteotomies are made. These osteotomies are outfractured and bent in place to allow significant expansion of the occipital and parietal regions of the skull. The barrel staves can be fixed in place by using cross struts of bone to hold the barrel staves out, preventing collapse by the soft tissue once the scalp is closed. The calvarial cap is then replaced and the height is reduced by gently displacing the cap inferiorly allowing the brain to expand posteriorly and laterally into the enlarged cranial vault. The cap is then fixed into its shortened position with sutures or absorbable plates and screws.

Midfacial Advancement

The next phase in the reconstruction of the child with a syndromic craniosynostosis is usually directed towards the midface depending on the growth and development of the craniofacial skeleton, the durability of the frontal-orbital advancement, and any pressing health concerns such as high ICP, airway obstruction, or severe exposure of the globes refractory to medical management. The appropriate operation focusing on the midface is usually a choice between a Le Fort III osteotomy and a monobloc procedure. Either of these procedures can be combined with an orbital bipartition to address hypertelorism. A monobloc operation allows the movement of the frontal-orbital bar, the malar midface, and the maxilla all as a unit, while the Le Fort III operation does not include the frontal-orbital bar. The monobloc requires an intracranial approach while the Le Fort III may not. The monobloc requires an osteotomy across the cranial base, creating an opening between the nasal cavity and the cranial vault, while the Le Fort III does not. A monobloc can be a more technically demanding operation and is felt to have a higher incidence of infection than a Le Fort III.

The combination of persistent intracranial dead space and an open connection to the nose is thought to contribute to the increased incidence of infection in the monobloc operation. A more selective approach to the monobloc procedure has been proposed to decrease the likelihood of infectious complications. Infants, patients with shunts, and older children with frontal sinus development have become relative contraindications to the monobloc procedure. Many surgeons think the aesthetic outcome of a monobloc procedure to be superior to other alternatives. All of these aspects of the procedures should be considered when contemplating a midfacial procedure on a syndromic patient.

The appropriate clinical indications for the choice of operation mainly reside in the clinical exam. A child with a recessed supra-orbital rim and frontal bone, apparent hypertelorism, malar hypoplasia and a class III malocclusion is a good candidate for a monobloc procedure. Alternatively, a repeat frontal-orbital advancement could be performed and followed by a Le Fort III several months later after complete healing. Still, another approach would be to perform a frontal-orbital advancement and a Le Fort III at the same time, which would leave the mid-portion of the cranial base and nasion untouched and therefore recessed. Such an approach would require camouflage onlay bone grafting to the nose but would also allow a differential advancement between the midface and frontal-orbital bar.

The appropriate age for midfacial advancement is controversial, advocates for early surgery point to the psychosocial impact of craniofacial disfigurement on the developing child, while proponents of delayed surgery discount those claims and point to the stability and the decreased number of operations to support their approach. At our institution, there have been no infectious complications when performing the monobloc procedure using the more selective criteria; therefore, the decision as to which procedure to use for midfacial advancement is purely made on clinical grounds.

Monobloc Advancement, Le Fort III Advancement, and Orbital Bipartition

The patient undergoing a monobloc advancement (Figs. 9 and 10) or a Le Fort III advancement (Figs. 11 and 12) requires a nasotracheal intubation, and the tube comes off along the cheek back to the anesthesiologist. The tube is sutured to the septum for security and the face and mouth are prepped completely. A methacrylate splint is made and fixed to the maxillary dentition with light gauge dental wire to stabilize the midface and maxilla in preparation for the instability after the bipartition.

The monobloc advancement requires a frontal craniotomy and the incision, and the dissection and exposure is similar to that described for the frontal-orbital advancement. In addition, the globes are degloved and the temporalis muscle is taken down in a similar fashion to the description for the frontal-orbital advancement; however, the zygomatic arch is dissected further medially and the orbits are dissected further laterally and inferiorly along the floor. The most superior osteotomy in the monobloc operation is made above and behind the superior orbital rim. A reciprocating saw is used to continue the cut along the orbital roof and then along the cranial base anterior to the cribiform plate and met by a symmetric osteotomy made on the opposite side. The lateral intra-orbital osteotomy is made using an osteotome or a reciprocating

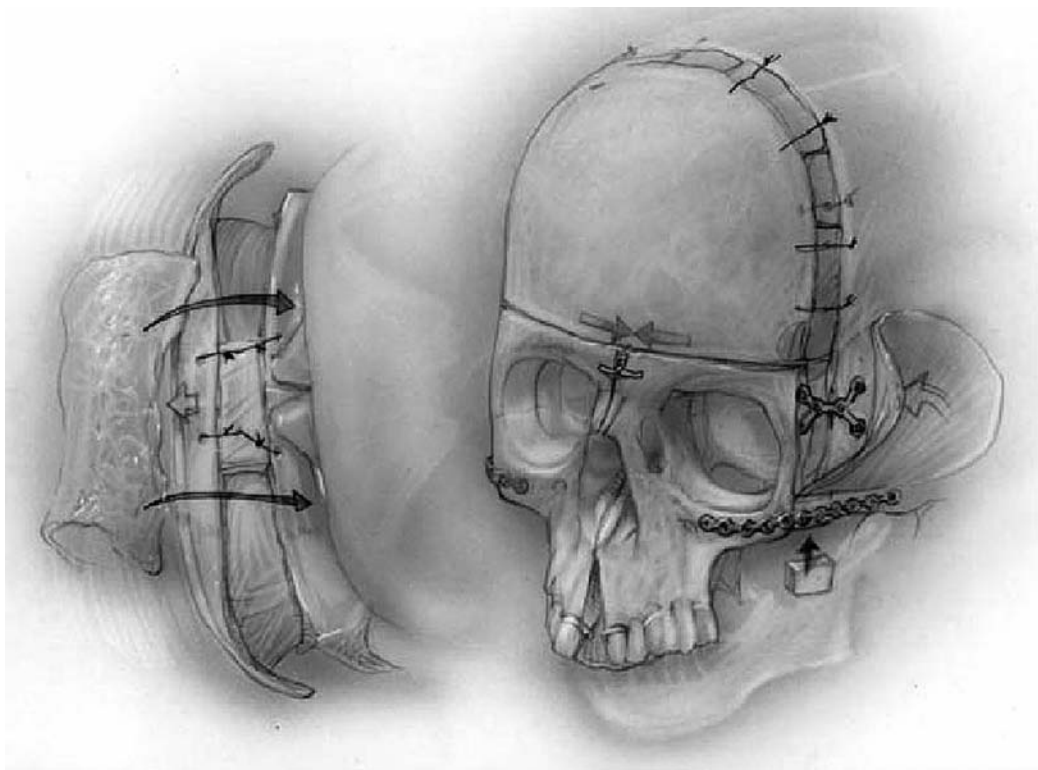


FIGURE 9 A schematic depiction of a monobloc advancement procedure.

saw and cut superiorly from the inferior orbital foramen to meet the osteotomy at the orbital roof. The orbital floor osteotomy is usually made with an osteotome and runs from the inferior orbital foramen in a line parallel to the inferior orbital rim but behind the posterior lacrimal crest into the substance of the laminae papyracea. The final intra-orbital osteotomy is performed with a 2 to 4 mm osteotome and runs from the medial orbital roof osteotomy on a line posteriorly to the medial canthus and medial to the globe ending in the substance of the laminae papyracea, completing the “donut osteotomy” around the globes. A straight zygomatic osteotomy is then performed in the thickest portion of the bone, allowing room for a stable plate to be fixed proximal to the osteotomy. An intra-oral pterygomaxillary disimpaction is then performed with a curved osteotome. Disimpaction forceps are then placed into the nose and secured against the maxillary palatal splint that was placed preoperatively. Carefully, the disimpaction forceps are moved forward and the perpendicular plate of the ethmoid is visualized through the cranial base osteotomy. A split osteotome is then used to cut the perpendicular plate, avoiding the nasotracheal tube. The disimpaction forceps are again thrust forward and also moved from side to side, dislodging the monobloc from the cranial base. It may be necessary to assist the disimpaction by placing a heavy periosteal elevator behind the maxillary tuberosity and pushing anteriorly, stretching the soft tissue restrictions. The monobloc should then be mobile and allowed to advance easily.

Once the monobloc is downfractured and released, the orbital bipartition can be performed to address hypertelorism. A marker is used to draw a “V” between the eyes with the bottom part of the V theoretically at the incisive foramen. In reality the V includes the mid-portion of the supra-orbital rim, the radix, and the central portion of the nasal bone. The width of the V will determine the extent of medial displacement attained. It is important to remove bone above and behind the medial canthus to allow the greatest correction; however, a stable medial canthal attachment to bone is crucial. The V is then cut and the bone is removed.

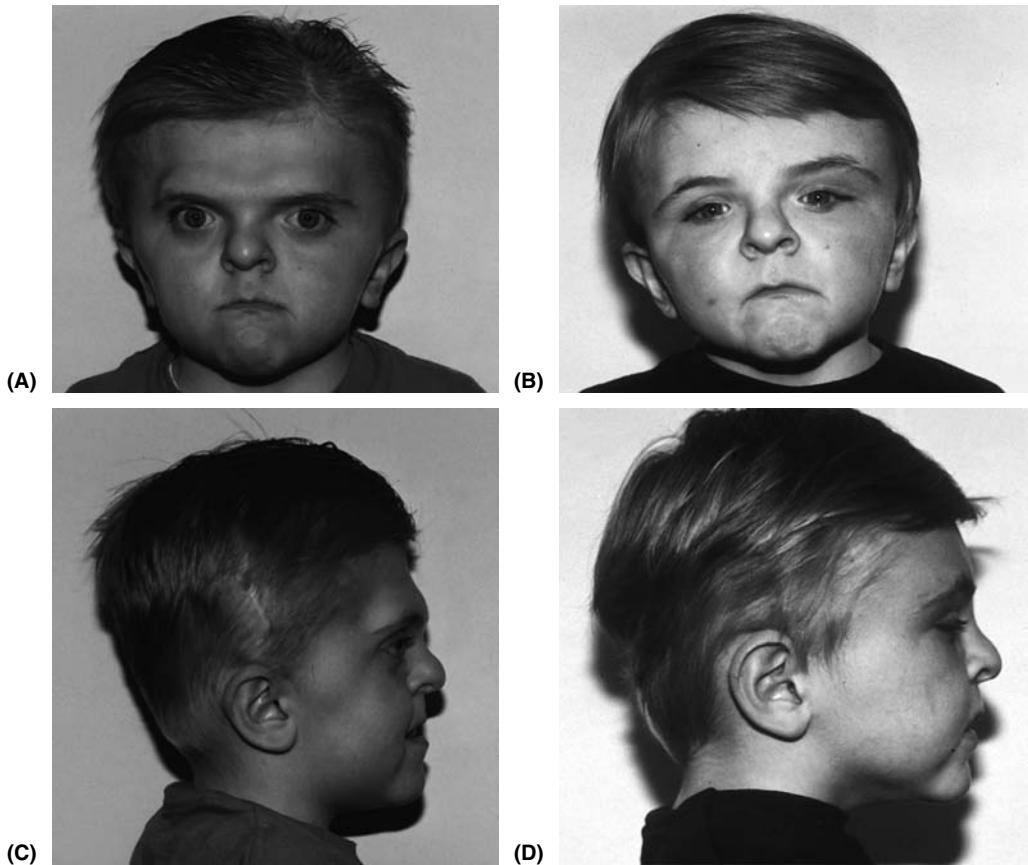


FIGURE 10 A child with Apert syndrome can be a wonderful candidate for a monobloc procedure. (A) Preoperative frontal view, (B) postoperative frontal view, (C) preoperative lateral view, and (D) postoperative lateral view.

Attention is then directed to the upper buccal sulcus where a small incision is made in the mucosa and a 2 mm osteotome is introduced between and behind the central incisors and carefully the midline palatal suture is separated. The use of a Lindemann burr to slot the bone carefully between the central incisors may facilitate the midline split. The intra-oral splint, placed earlier, is now functioning to stabilize the two segments of the face. A drill hole is then made into the stable bone lateral to the V cut on both sides; a strong wire is placed through both holes and twisted down. The medial canthus and medial orbits are drawn toward each other, and the maxillary plane is somewhat flattened. After completing the orbital bipartition the monobloc can then be advanced and the lateral portions can be “bent” to allow a transverse convexity to the face as desired. The malar region is a key landmark to the appropriate degree of advancement, but in the end the advancement and reposition of the monobloc is based on clinical judgment. It is important to register the monobloc vertically so as not to cause a significant open bite. Once the correct position is ascertained, a rigid 2.0 plating system is fixed at the zygomas bilaterally giving the advancement the greatest amount of stability. Additional plating behind the orbital rim, usually a 1.0 or 1.5 plating system is used to reinforce the fixation. Interpositional bone grafts are then placed in the open space behind the advanced edges of the bone and “lagged” to the plates.

After fixing the monobloc into an advanced position, attention is directed to the intracranial opening in the cranial base. The intranasal mucosa should be visible and if possible can be stripped, mobilized, and closed. A detailed, form fit, piece of bone graft is then cut and placed between the edges of the bone, filling the void completely, and reestablishing

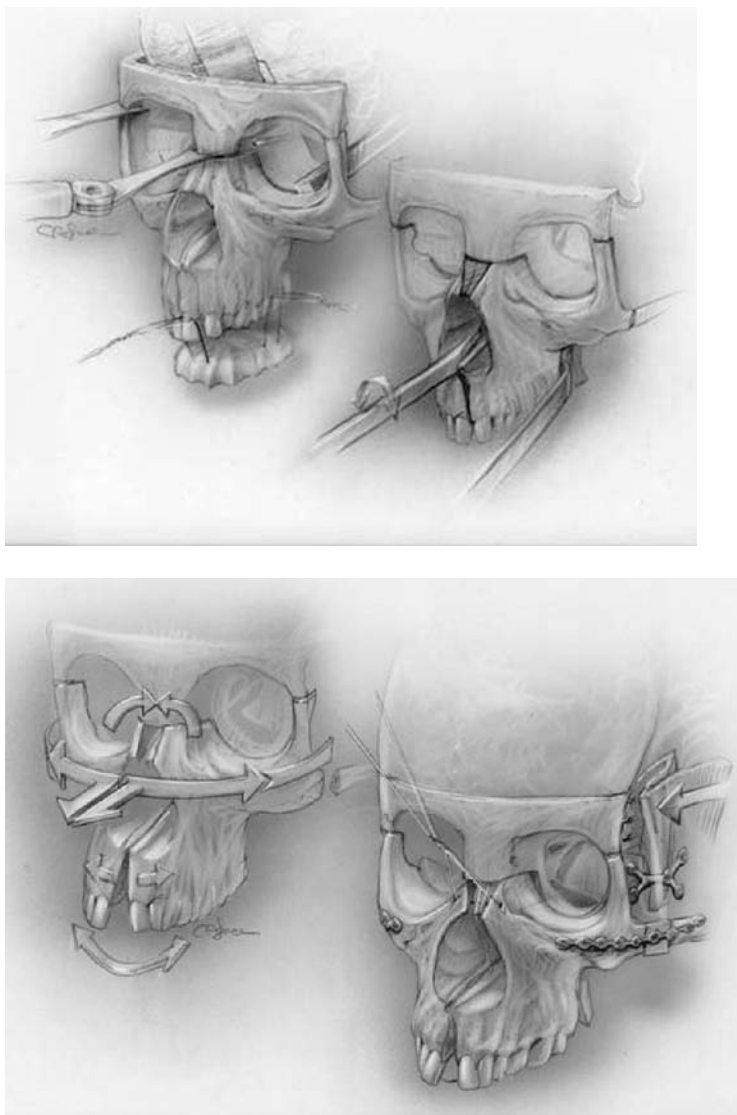


FIGURE 11 A schematic depiction of a Le Fort III advancement procedure.

a separation between the nasal cavity and the cranial cavity. A piece of pericranium is placed above the bone graft, and sewn to surrounding periosteum as an insurance layer, reinforcing the separation. The frontal craniotomy is then reshaped as necessary and attached to the advanced frontal-orbital rim using 1.0 microscrews and plates. All of the defects in the cranial cavity need to be closed with grafts, as the osteogenic potential of the dura are minimal at this age. The coronal flap is then closed in a similar fashion as described for the frontal-orbital advancement. The patient is sent to the ICU for monitoring, and may be ready for extubation the next day. Some surgeons advocate leaving the child intubated for a few days to avoid positive pressure in the nose, and allow the bone graft at the cranial base to “stick.” The most important aspect of extubation is to avoid bagging the patient with a facemask, which does impart this positive pressure and can damage the cranial base reconstruction. The hospital course for this operation may be 5 to 10 days.

A Le Fort III (Figs. 11 and 12) is very similar to a monobloc procedure (Figs. 9 and 10), but avoids the cranial base osteotomy. The lateral cuts at the orbital rim are made lower and



FIGURE 12 This child with Crouzon syndrome underwent a Le Fort III midfacial reconstruction. **(A)** Preoperative oblique view, **(B)** postoperative oblique view, **(C)** preoperative lateral view, and **(D)** postoperative lateral view.

the midline nasal cut is made across the nose below the cranial base. A coronal CT scan can guide the surgeon as to the level of the cranial base. If the cranial base is high, the Le Fort III can be done extracranially by using the anterior ethmoidal artery foramen as a landmark and safely cutting below this level at the midline. If the cranial base is low, or the surgeon is uncomfortable with the "blind" cut, an intracranial approach will give better visualization and allow a safe midline osteotomy. The rest of the Le Fort procedure is completed in a similar fashion as the monobloc except that the cranial base does not require reconstruction as there is no defect created. Since the V cannot be as wide in a Le Fort III, because it is done at a lower level, the degree of correction allowed in a concomitant orbital bipartition is decreased. Finally, the degree of advancement at the lateral orbital rim is dictated by the degree of projection of the stable forehead and supra-orbital rim, a bony step should be avoided and a smooth transition established. A lateral canthopexy is often performed in both the monobloc and the Le Fort III operation and done in a similar fashion as outlined in the section on frontal-orbital advancement.

The monobloc and Le Fort III operations are not meant to gain normal occlusion; indeed the patient is in mixed dentition and future growth of the jaws precludes normalization at such an early age. The periorbital and malar areas should be the landmarks used to guide the correction of the midface, this should be done while advancing the maxillary occlusion as much as possible but without sacrificing midfacial appearance. An orthognathic procedure, such as Le Fort I with or without a mandibular osteotomy, is the rule and not the exception. The orthognathic procedure is done after skeletal maturity is complete and mandibular growth is concluded. Close communication with the orthodontist is mandatory as the orthognathic procedure is the last main osteotomy required. A Le Fort I osteotomy is usually required to correct the persistent class III malocclusion that exists at skeletal maturity in these patients. In the case of large occlusal discrepancies, a bilateral sagittal split osteotomy of the mandible can bring about a normal occlusion, give a more salutary appearance to patient's facial proportions, and decrease the incidence of relapse associated with significantly large advancements of one jaw. A genioplasty can be a very effective procedure when combined with upper or lower jaw orthognathic surgery to bring about an improvement in facial proportion and a more aesthetic appearance when reconstructing the syndromic patient. These orthognathic procedures are performed with the same indications and planning in the syndromic patient as in the nonsyndromic patient and are therefore not outlined here.

Midfacial Distraction Osteogenesis

Although distraction osteogenesis of the midface for monobloc or Le Fort III operations is a relatively new field, it may have its most appropriate applications in cases where conventional surgery is inadequate. Such an example can be found in the child below four to five years of age that shows signs of increased ICP despite shunting, exorbitism threatening globe exposure, and sleep apnea secondary to severe midfacial retrusion. Patients with all or some of these symptoms and signs pose a difficult management problem as they require cranial vault expansion as well as midfacial advancement, but have contraindications to the monobloc procedure (young age, and a shunt in place). Distraction osteogenesis could allow the appropriate cuts to be made in a monobloc procedure but would avoid a true defect in the cranial base. A callous would form at the osteotomy site and would be stretched slowly to the expanded length and theoretically avoid the problem of a contaminated enlarged dead space at the cranial base. Utilizing the same reasoning a distraction osteogenesis monobloc procedure has been performed successfully for these same indications at our institution (Fig. 13).

Children with syndromic craniosynostosis require life long follow up for a variety of issues. The growing child is in a dynamic state. There can be a mismatch of brain growth versus skull growth, which can lead to intracranial crowding and may promote the development of a Chiari I malformation or cerebellar tonsillar ectopia. Regular neurological assessment is important to assure that such problems are quickly addressed. The development of headaches in such children may indicate intracranial crowding. Other symptoms may be more subtle such as a decrease in school performance or coordination, change in vision or loss of milestones. Careful radiological evaluation and detailed neurological assessment are important. We advise that all children with craniofacial anomalies under go regular ophthalmological evaluation, which includes dilated fundoscopic examination. Unmonitored increases in ICP can be

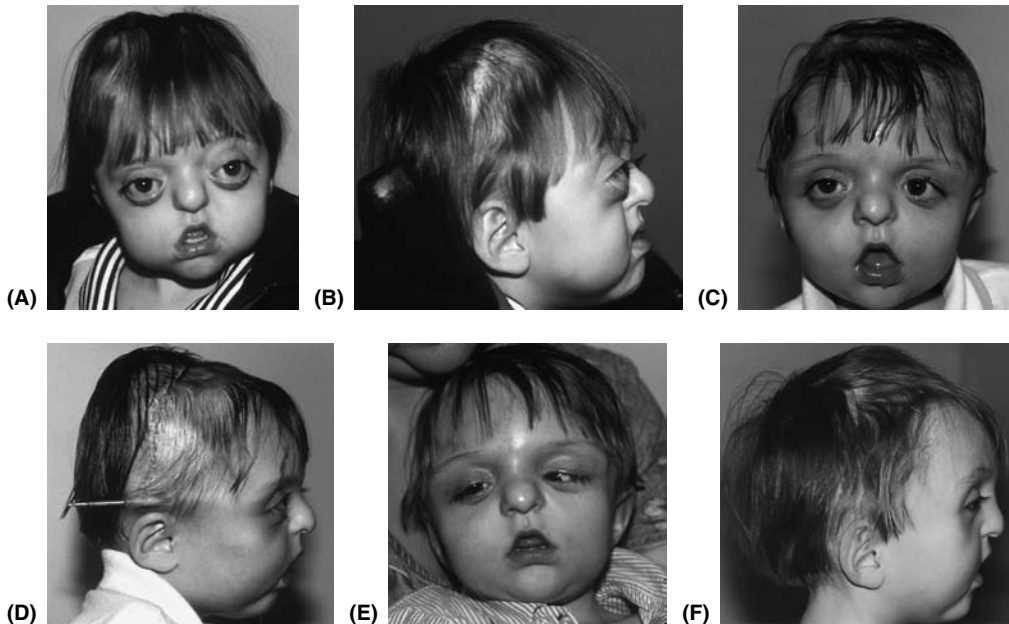


FIGURE 13 A monobloc advancement procedure using distraction osteogenesis in a two year old with Pfeiffer syndrome showing signs of increased intracranial pressure, exorbitism, and sleep apnea. (A) Preoperative frontal view, (B) preoperative lateral view, (C) postoperative frontal view during the distraction period, (D) postoperative lateral view during the distraction period, (E) postoperative overcorrected frontal view after completion of distraction, and (F) postoperative overcorrected lateral view after completion of distraction.

devastating, leading to permanent neurological deficits such as blindness. In cases in which intracranial crowding is not definite, but suspected, the use of an ICP monitor can sometimes be useful. It can help sort out whether there is any evidence of increased ICP or whether symptoms such as headache are better managed medically.

The care of the syndromic patient with craniosynostosis is perhaps the most challenging problem in craniofacial surgery. The syndromes vary in both their presentation and severity; however, many of the structural and functional sequelae have common characteristics. The multifaceted medical problems of these children are particularly well suited to a multidisciplinary team approach to their care. The surgeons must be aware of the most pressing health concerns of these patients and incorporate those concerns into a well-formulated phased plan of comprehensive reconstruction. The overall goals of treatment should continually guide the care of these exceptional children; to provide a predictable and durable reconstruction that will restore facial form, reestablish symmetry, attain the best results in the least number of operations, and infuse the patient with a sense of both emotional and physical health benefits.

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8 Successful Separation of Craniopagus Conjoined Twins Using a Staged Approach: An Evolution in Thought

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INTRODUCTION

Conjoined twins are a rare entity that presents very special challenges to the medical and surgical teams that care for them. The separation of craniopagus twins is an even more extreme and rare challenge. Craniofacial surgery requires a coordinated effort between plastic surgery and neurosurgery, and it is clear that separating craniopagus twins takes this coordination and cooperation to a stratospheric level. As advances in medicine have taken place, the risk of separating craniopagus twins remains daunting with survival ranging about 50%. The true incidence of craniopagus and the outcome of separation around the world are not known because of incomplete reporting, but frequently these cases are reported in the media as curiosities. While outcome is affected by the extent of shared tissue, in those cases where shared brain vasculature is included, the ultimate goal, which is to have both twins emerge from their separation with full neurological function and a chance at leading independent and productive lives, has remained elusive.

In March 2003, we were contacted to evaluate craniopagus conjoined male infant twins for possible separation. In order to assess this possibility, we reviewed the literature. Briefly, this review along with personal interviews revealed complications that included death of one or both twins, brain exposure, meningitis, and neurological compromise. The most common approach included a preliminary procedure to place tissue expanders under the scalp followed by a surgical separation performed over many hours. We hypothesized that the morbidity and mortality were primarily due to the prolonged surgery needed to separate such twins, and inadequate time to allow for vascular adaptation. In order to minimize morbidity and mortality, and preserve function, we designed an open-ended, multi-staged separation in order to allow them to improve their venous collateral circulation, and recover from each stage before progressing to the next stage. Four major stages over nine and a half months led to their successful separation and preservation of neurological function. To our knowledge, this is the first time an outcome like this was achieved in such a case.

A review of the pertinent literature, as well as our rationale and methodology are discussed in this chapter.

Craniopagus twins occur in about 2% of conjoined twins. In craniopagus, the union can occur anywhere on the cranial vault but by definition, does not involve the foramen magnum, face, vertebrae, or skull base. The thorax and abdomen are completely separate. The junction of the conjoined twins is rarely symmetric and can involve any part of the meninges, venous sinuses, and the cortex. Axial and rotational orientation is variable. Each of these factors can influence the development, distortion, deformation, and displacement of the brain, the meninges, and the vascular system as well as the prognosis after surgical separation.

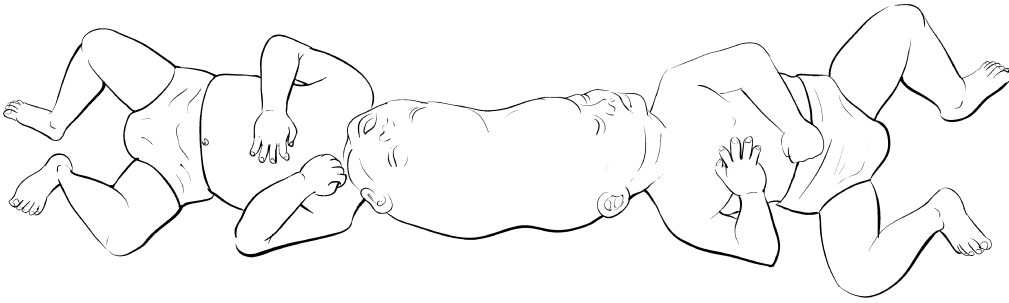


FIGURE 1 Craniopagus twins. *Source:* From Medical Modeling LLC, Golden, Colorado, U.S.A.

There are four types of conjoined twins joined at the head: (i) *Craniopagus* are joined only in the calvarium (Fig. 1); (ii) *Cephalopagus* are joined ventrally, from the top of the head down to the umbilicus (Fig. 2); (iii) *Parapagus diprosopus* are joined laterally with two faces on one head but share only one body (Fig. 3); (iv) *Rachipagus* are joined dorsally along the vertebral column (Fig. 4), occasionally involving the occiput.

The female to male ratio in craniopagus twins is about 4:1 which is similar to the ratio for other types of conjoined twins. Other reported anomalies include congenital heart disease, cleft lip, cleft palate, supernumerary thumbs, extrophy of the cloaca, and absence of the entire urinary tract, bladder extrophy, absence of anus and vagina, and imperforate anus (1).



FIGURE 2 Cephalopagus twins. *Source:* From Medical Modeling LLC, Golden, Colorado, U.S.A.

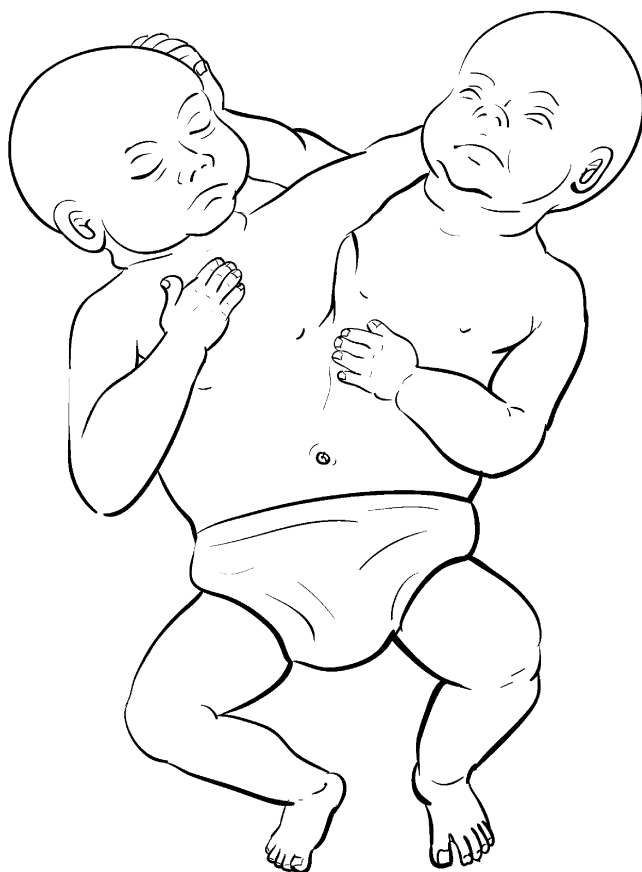


FIGURE 3 Parapagus twins. *Source:* From Medical Modeling LLC, Golden, Colorado, U.S.A.

CLASSIFICATION

The simplest method of classification of craniopagus twins makes reference to the site of junction: *frontal*, *temporal*, *parietal*, or *occipital*, and combinations of them. O'Connell has suggested the use of the terms *partial* and *total* to describe the degree of junction (1). He further describes types I, II, and III to indicate whether the twins are facing the same or opposite directions or their axes were perpendicular to each other. Our case represents an O'Connell type I (Fig. 1).

Winston et al. described a classification system based on the embryological origin of the deepest shared structure: the surface ectoderm and cranium; the dura (ectomeninx); the leptomeninges (endomeninx); and the neuroectoderm (2). Todorov et al. correlated postoperative survival with the axial orientation of the union. He describes the acute frontal–frontal angle was most favorable, and mortality increased as that angle increased (3).

Spencer points out that each of these systems has merit, but they fail to take into account the anatomy of the underlying dural venous sinuses. She points out that it is the specific anatomy of these vessels that “doom the vast majority of these infants to the wretched life of intact conjoined twins or the significant risk of death or serious neurologic impairment following surgical separation” (4).

The pathological configuration of the dural sinuses appears to be influenced by: (i) the site of union; (ii) the plane of intersection, wherein the larger the plane of intersection, the greater the probability that the sinuses and cortical surfaces are fused; and (iii) the rotation and angulation from the frontal area. It also follows that the larger the area of confluence of the scalps, the greater the soft tissue requirement for reconstruction.

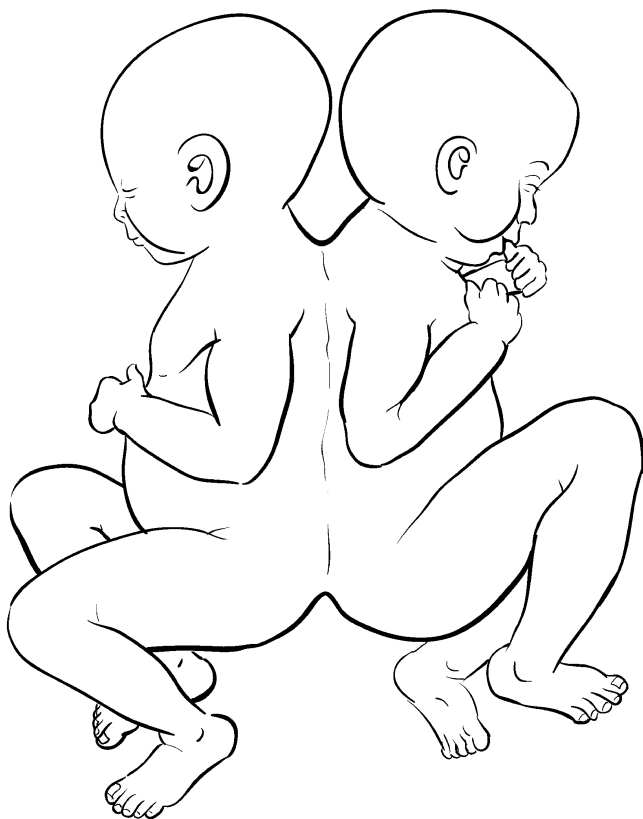


FIGURE 4 Rachipagus twins. *Source:* From Montefiore Medical Center, LLC, Golden, Colorado, U.S.A.

The dural sinuses are not true veins; they do not contain valves, muscles, or fibrous walls and are therefore very fragile. The sinuses are not movable within the surrounding tissue. The location of the sinuses in the conjoined area may be hard to predict, and the direction of blood flow is unpredictable. When the twins are joined vertex-to-vertex, the falx cerebri does not form and subsequently the superior sagittal sinus cannot develop. The analog of the superior sagittal sinus forms around the periphery of the conjoined plane in a fold of dura. This shared dural hoop contains a venous sinus that can be either completely or partially circumferential. This may also form a “venous lake.” Each twin will have veins draining into, or out of, this sinus. Extracranial arteries are an important consideration when planning skin flaps but it is rare that the intracranial arteries are involved.

Winston points out that adequate support must be provided to avoid gravitational forces on the brains during separation surgery. The possibility of air embolism is of concern during separation as well (2). Any external pressure on the jugular veins must be carefully avoided since this can increase venous pressure, and subsequently bleeding and cerebral edema. Because valves are not present in the dural sinuses, the relative position of the twins on induction of anesthesia and surgery may cause some degree of “transfusion” from one twin to the other. The issue of adrenal dominance is important in any kind of conjoined twinning; the twin with adrenal suppression may require steroid supplementation preoperatively, intraoperatively, and postoperatively (5).

HISTORICAL EXPERIENCE WITH CRANIOPAGUS CONJOINED TWINS

The first attempt at separation of craniopagus recorded in the twentieth century involved 12-day-old infants with parietal union; neither survived this attempt (6). The first survival recorded was of 14-month-old males with a large parietal union, a shared sinus in

a semicircular dural shelf, and a single confluence. There was massive hemorrhage when the veins were divided. The first twin died 34 days after surgery, but the second survived with a temporary hemiparesis but died from complications of hydrocephalus at 11 years of age (7). The first recorded survival of both twins was in a set of seven-month-old girls with minimal parietal union. The authors describe a thin sheet of bone across the plane of union. They shared a 5 mm segment of the superior sagittal sinus. One twin was reported to have survived neurologically intact in spite of massive hemorrhage, but the other suffered severe neurologic injury. Interestingly, the neurologically impaired twin donated a kidney to her twin sister 28 years later (4,8).

It is interesting to note that efforts have been made to alter the size and angulation of the union. Wolfowitz et al. successfully increased the angle of union between a set of frontoparietal twins in an effort to increase the surgical exposure (9). In another case, a metal band was placed circumferentially around the plane of junction, but had required removal when the twins developed seizures (10). In yet another set of craniopagus conjoined twins, constriction by a plastic ring was abandoned when the resulting ulcer became infected (11). An adjustable pneumatic cuff was placed around another set and the bridge was decreased by 10% and prevented subsequent growth of the conjoined area, but the five-month-old males with a minimal occipito-parietal union died of massive hemorrhage in the operating room (12). In a German case with extensive parietal union, a circular nylon band was used unsuccessfully (13).

In her review on this topic, Spencer notes that the most important data to be obtained prior to surgery concerns the location and character of the dural sinuses (4). It is clear that division of the veins draining into the dural sinus can result in elevation of the cerebral venous pressure and venous infarcts. In addition, such elevation in venous pressure may also result in significant cerebral edema making reconstruction more difficult. Cardiopulmonary bypass, hypothermia, intraluminal shunting, and circulatory arrest with division and reconstruction of the major sinuses has led to minimal success (14).

A craniopagus was separated in stages with gradual closure of a bridging vein. To accomplish this, a screw clamp with an exteriorized stem was used over several days to gradually occlude a short bridging vein between superior sagittal sinuses; this method led to a successful separation but no long-term follow-up was reported (15).

Continued advances in anesthesia techniques as well as critical care has improved the survival of craniopagus separation. Kawamoto and Lazareff inserted tissue expanders and then followed tissue expansion with a single-stage separation of a set of Guatemalan female craniopagus twins. A similar technique was used by Salyer and Swift to separate a pair of male twins from Egypt. In the latter set a rotating table was fabricated to allow the twins to be rotated along their axis in unison. In these cases, each twin survived, but there were notable neurologic deficits more than six months after each separation.

A set of 29-year-old Iranian women joined temporoparietally suffered fatal hemorrhage during their attempted separation. While medical models were utilized, and the surgeons felt that they had the advantage of many recent technological innovations, they noted that the final bridging vein, after hours of surgery to divide the other veins, was enormously dilated and there was significant venous engorgement. This engorgement is likely due to the venous hypertension from the abrupt change in venous pathways.

AN EVOLUTION OF THOUGHT

A careful review of personal communications and the literature have convinced us that a lengthy operation to ligate and divide the bridging veins in such cases leads to unavoidable venous hypertension and circulatory compromise of the brain. Plastic surgical principles naturally lead one to consider staged division of venous tributaries to encourage the dilatation of venous collaterals and the resulting diversion of drainage (16).

While previous staged techniques have been attempted with mixed results, we feel that the stages must be timed such that complete recovery is allowed to ensue prior to subsequent stages. These breaks between surgical stages would allow for occupational and physical therapy and nutritional support and would allow time for the desired vascular changes to

occur. The intent of the staged approach is that it would allow the patients to be exposed to general anesthetics for shorter periods of time, require less intravenous fluids, have less bleeding, less cerebral edema, and require less transfusion products. Less cerebral edema would allow for easier and more reliable soft tissue coverage at the completion of their separation. While the skin flaps must be completely designed from the beginning of the staged procedures, elevating the flaps for each stage would offer the delay phenomenon to the skin flaps. Perhaps the most important benefit of the staged procedure is that it allows the bridging veins to be divided sequentially, thereby allowing collateral venous drainage to improve. We have hypothesized that the improved venous drainage, together with the previously discussed advantages, would improve survival as well as the neurologic outcome. At the final stage, during separation, improved venous drainage, and shorter surgical time prior to separation would cause less cerebral edema, thereby making dura and scalp reconstruction less complicated (17,18).

CASE REVIEW

On April 21, 2002, a 29 year-old woman in the Philippines gave birth to a set of male craniopagus twins by Ceasarian section. A level-2 ultrasound had diagnosed the condition, and termination of the pregnancy was offered but rejected because of religious conviction. She desired separation for them and a work-up was obtained (Fig. 5).

During intubation for radiographic studies, twin A suffered aspiration pneumonia twice and further efforts at work-up were abandoned and transfer to the Children's Hospital at Montefiore was arranged. Arrangements were made to augment their nutrition, provide pulmonary care, and intensive occupational and physical therapy at Blythedale Children's Hospital in Valhalla, New York. We have had a long-standing clinical relationship with Blythedale, which is a rehabilitation center that does not provide acute care.

Upon arrival in New York, evidence of failure to thrive was present, and twin A, much smaller than twin B, was noted to have severe hypertension. Antihypertensive medications were prescribed. Urine output was also noted to be greater in twin A.

Craniofacial computed tomography (CT) with three-dimensional reconstruction was obtained as well as magnetic resonance imaging/magnetic resonance venography (MRI/MRV). Data from these studies were used to obtain diagrams and clear stereolithographic models (Medical Modeling, LLC, Golden, Colarado, U.S.A.) of the shared venous system within the conjoined skulls (Fig. 6). In addition, a three-dimensional model of the skin envelope was also obtained (Medical Modeling, LLC, Golden, Colarado, U.S.A.). This model is quite useful to the plastic surgeon and a pencil can be used to design skin flaps (Fig. 7A,B).

Scalp flaps were designed to maximize their blood supply and avoid suture lines over the vertex of the scalp once the twins were separated. A sinusoidal pattern was designed to



FIGURE 5 Craniopagus twins referred for separation.



FIGURE 6 Illustration showing skin envelope with shared venous sinus system superimposed from initial computed tomography data; the lesser vessels are eliminated for clarity. Twin A on right. *Source:* From Montefiore Medical Center/Medical Modeling, LLC, Golden, Colorado, U.S.A.

accomplish this goal. While opposing pericranial flaps were considered, they were not used because of the difficulty in incorporating this design into a multi-staged procedure.

Flaps were designed to extend toward the opposite twin's right ear, leaving the incisions to resemble a sinusoidal curve around the conjoined scalp. Because the twins' interaxial angle was slightly less than 180° (Fig. 7), one twin would have a shorter flap. While tissue expansion would be utilized, the twin anticipated having the larger dura defect and patch was given the longer flap. It was crucial to identify this prior to the first stage of separation.

Craniofacial CT and MRI/MRV studies were also used to generate holograms (Voxel, Inc., Provo, Utah, U.S.A.). To our knowledge, this is the first time holographic images have been used in the separation of conjoined twins (Fig. 8A,B).

If a single-staged separation of craniopagus twins were being planned, tissue expansion could be performed on the intact scalp without previous surgery, thereby minimizing the risks associated with tissue expansion (e.g., exposure and infection) (19). During the separation, however, the expanders require removal, and the expanded scalp undergoes a degree of contraction during the hours of neurosurgery required to separate the twins in a single-staged separation. Tissue expansion during a multiple-staged separation, however, is more complicated. The need to perform tissue expansion after previous stages, with healing scars and manipulated tissue is a challenge unique to the staged approach. In each of the stages, exposure is provided through the previously designed flaps, and the craniotomies are kept as narrow as

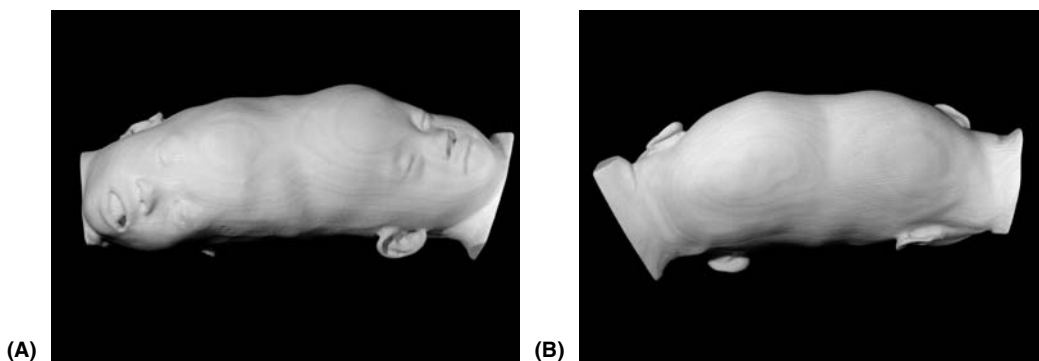


FIGURE 7 Three-dimensional model fabricated from computed tomography data. (A) Anterior view; twin A on left. (B) Posterior view; twin A on right. *Source:* From Montefiore Medical Center/Medical Modeling, LLC, Golden, Colorado, U.S.A.

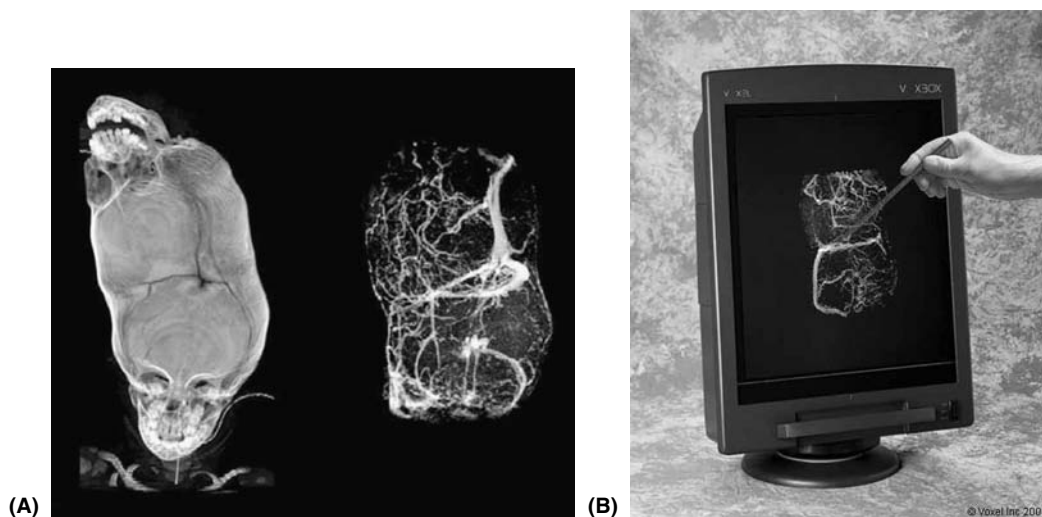


FIGURE 8 (A) Hologram from computed tomography data on left and magnetic resonance venography data on right (these can be overlaid). Twin A is at the top. (B) Holograms projected in front of viewbox. *Source:* From Montefiore Medical Center/Voxel, Inc., Provo, Utah, U.S.A.

possible along the conjoined perimeter. Originally, tissue expansion was planned for each stage, replacing the expanders at each surgery with a larger size. We felt that gradually increasing the size of the expanders would be needed, as the healing scars grew longer with each stage.

Multiple dental caries were noted on examination. These represented possible sources of bacteremia, and multiple extractions were performed under general anesthesia. Anesthesiologists were, from that point forward, dedicated to the continued care of each specific twin throughout their various stages. Our dedicated craniofacial surgery scrub technicians and circulating nurses made the same commitment.

Stage 1

On October 20, 2003 stage 1 was performed. Intraoperative navigation was utilized (Stryker). The conjoined twins were positioned supine and the forehead was opened along the planned incision line exposing the shared frontal bone. A frontal craniotomy was performed and dura was divided adjacent to the line of fusion on the side of twin B. A circumferential “shelf” of dura was seen. This shelf was the dura enveloping the circumferential sinus, which would ultimately be left with twin A. Brains were seen to be separate. A large anterior bridging vein in twin B was divided at this stage (Fig. 9). A silastic sheet was placed between the brains to avoid adhesions. Dura was closed, and the craniotomy was closed with titanium miniplates. We typically use resorbable fixation in our pediatric craniofacial work, but because hardware would need to be removed during the final separation, titanium hardware was applied. 100 cc tissue expanders (Inamed, Santa Barbara, California) were then placed adjacent to the craniotomy and the scalp was closed in layers.

Subgaleal fluid leaked from the suturelines 10 days postoperatively and worsening fevers led to removal of the tissue expanders. Otherwise, recovery was uncomplicated.

Stage 2

On November 24, 2003, the twins returned to the operating room for the second stage. The skin incision was lengthened toward the left ear of twin A and further to the occipital area, allowing for the second craniotomy to be performed. Bridging veins were temporarily clamped on twin B's side of the circumferential sinus. No sign of cortical edema was observed so these veins

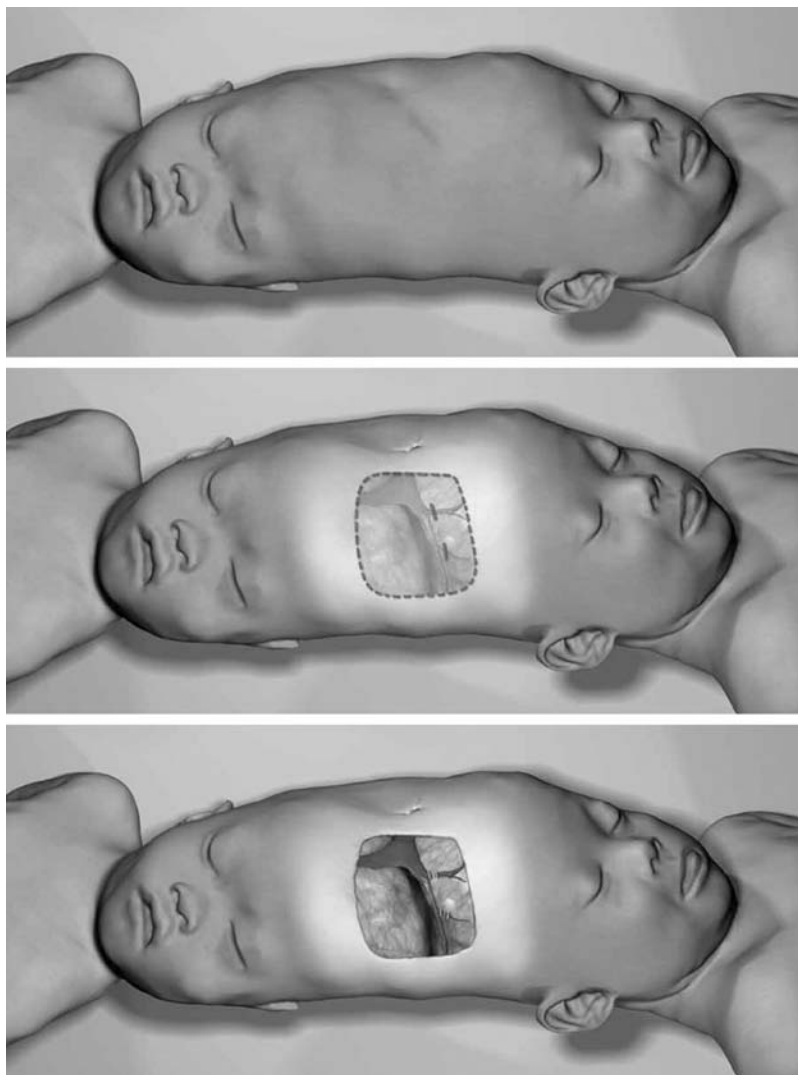


FIGURE 9 Note orientation and large bridging vein that was ligated (clipped) and divided, leaving the circumferential venous sinus with twin A on left. *Source:* From Montefiore Medical Center/Medical Modeling, LLC, Golden, Colorado, U.S.A.

were ligated and divided (Fig. 10). The previously placed silastic sheet was replaced. Closure was performed similarly after placement of new tissue expanders. The tissue expanders were again removed 14 days later for similar reasons as the prior stage. At this point, we felt that tissue expansion should be performed separate from the craniotomies because the usual cerebrospinal fluid leaks into the surgical site.

Stage 3

New CT scan and MRI/MRV were obtained and on February 20, 2004 the third stage of their vascular separation was performed. CT and MRI/MRV studies were repeated between stages to confirm the development of the needed venous collaterals (Fig. 11). From the first procedure, timing between stages was subjectively judged with regard to their progress in physical therapy as well as their soft tissue healing. No predetermined schedule was utilized.

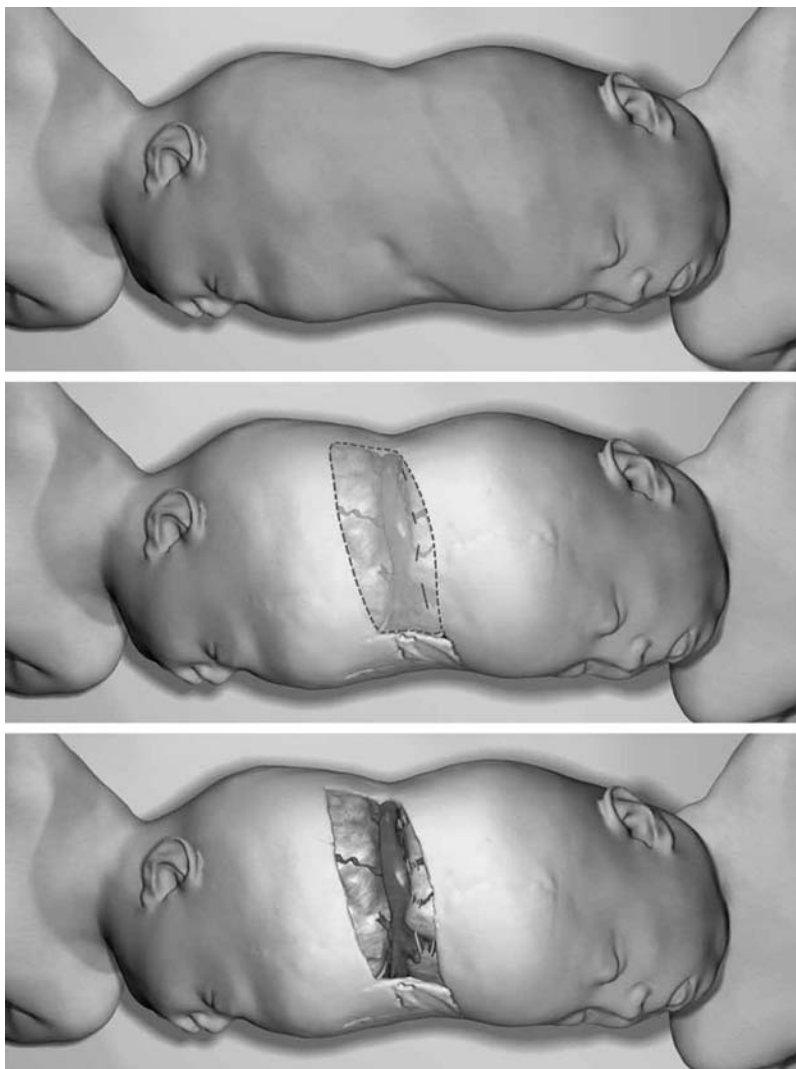


FIGURE 10 Note orientation and bridging veins that were ligated (clipped) and divided, leaving the circumferential venous sinus with twin A on left. *Source:* From Montefiore Medical Center/Medical Modeling, LLC, Golden, Colorado, U.S.A.

The twins were placed in prone position and the incision was lengthened along its pre-designed path and an occipital craniotomy was performed. Vessels were addressed in similar fashion and closure was performed (Fig. 12).

Tissue expanders were placed eight weeks prior to their expected separation surgery. Expanders were placed over each left ear, through separate incisions behind the ears, in areas that were not previously operated upon. Subcutaneous ports were placed behind the earlobes, and expansion was begun three weeks after placement.

Stage 4

Final separation surgery was performed on August 4, 2004, nine and a half months after the first stage. Placement of the tissue expanders above the ears allowed them to remain in place while the separation was performed. Continued circumferential dissection and division of the remaining dura and veins allowed their separation in a controlled fashion. In spite of abundant

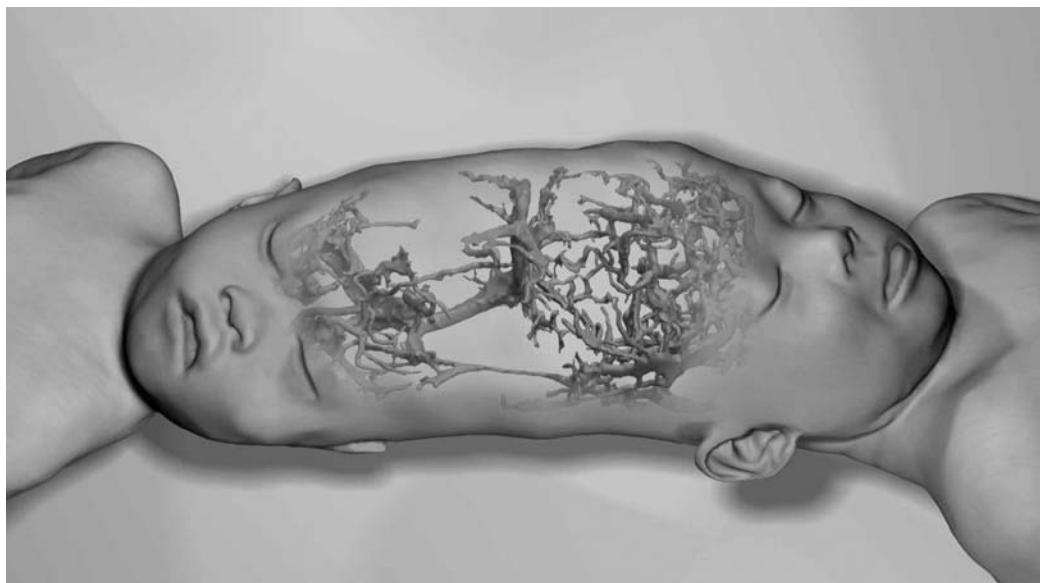


FIGURE 11 Increased quality of venous collaterals; twin B on right. *Source:* From Montefiore Medical Center/Medical Modeling, LLC, Golden, Colorado, U.S.A.

preoperative imaging, a shared posterior temporal lobe was encountered and divided according to its vascular pattern (Fig. 13).

Once the twins were separated, the operating tables were rotated approximately 30° to allow teams to work side-by-side. This rotation was felt to require the least amount of movement in the operating room, (tables pivoted on their pedestals and anesthesia teams did not need to move) and was therefore felt to be the safest maneuver. Durasis® (Cook Biotech, West Lafayette, Indiana, U.S.A.) was used to replace missing dura, tissue expanders were removed and scalp flaps closed. Twin B underwent completely primary closure over the large dural graft, while Alloderm® (LifeCell, Branchburg, New Jersey, U.S.A.) was placed over the native dura over the right ear of twin A.

Through all stages of surgery, the twins shared 4175 cc of packed red blood cells. No drains or primary shunts were used during the final separation, nor were they required subsequently. The twins were lightly sedated until their extubation of postoperative day 3 for twin B and postoperative day 4 for twin A. Prophylactic phenobarbital was used for each procedure. Twin A's antihypertensive medications were discontinued one week after surgery. Neither twin has developed hydrocephalus. Wounds have healed without cerebrospinal fluid leak (Fig. 14). In twin A, split thickness skin grafts were placed over the Alloderm three months after separation.

At the time of this writing, 32 months after separation, both twins are neurologically intact, interactive and playful; they currently feed themselves. Each twin walks without the aid of walkers. Since their separation they required tonsillectomy and adenoidectomy, as well as myringotomies for chronic otitis media. They have each developed speech and continue to gain the developmental milestones that were delayed by the nature of their conjoined status.

As healing progresses, additional scalp flap advancement, with or without tissue expansion, will provide complete scalp coverage in twin A; calvarial vault reconstruction will follow in each twin (Figs. 14–16). We are delaying reconstruction in order to allow them as much time as possible in school, therapy, and as a family. Fourteen months after separation, a three-bedroom home in the community was made available to the twins and their mother. The twins commute daily by school bus to therapy and school. There are no detectable neurologic deficits according to our neurology colleagues.

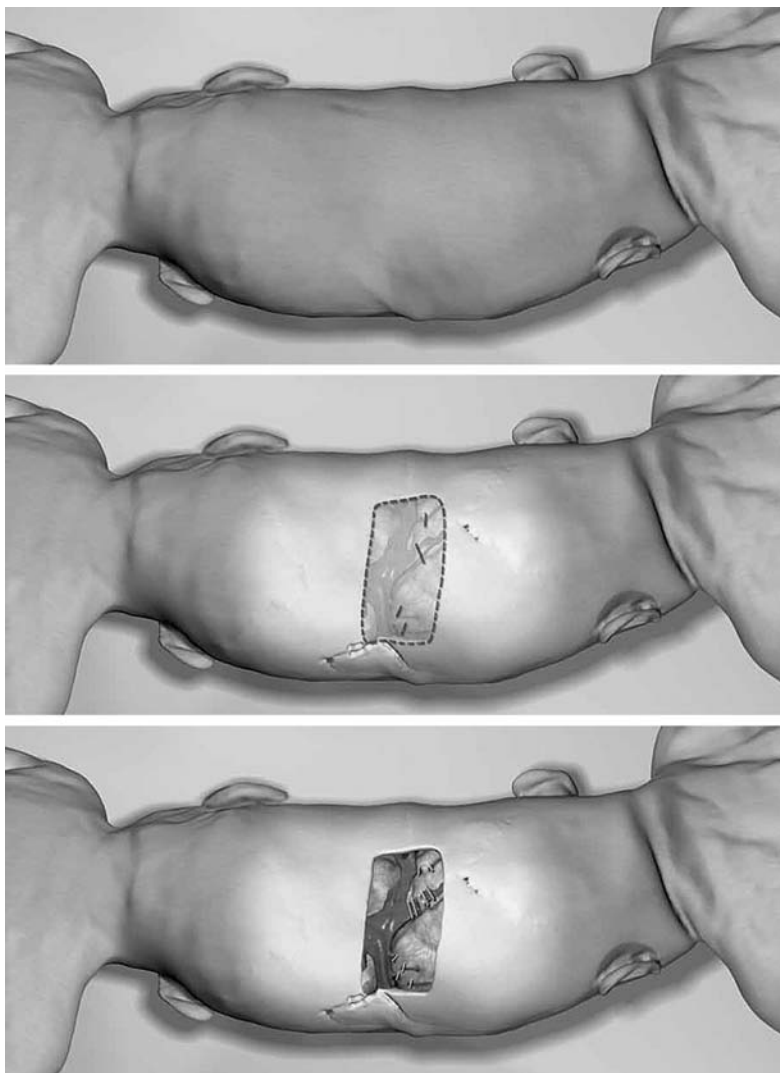


FIGURE 12 Note orientation and bridging veins that were ligated (clipped) and divided, leaving the circumferential venous sinus with twin A on left. *Source:* From Montefiore Medical Center/Medical Modeling, LLC, Golden, Colorado, U.S.A.

DISCUSSION

Craniopagus is a very rare entity. While advances in anesthesia and critical care management have been significant, survival from separation surgery remains about 50% over the past decades. Among the surviving twins however, neurological injury has been common in reported cases of O'Connell type I twins. To our knowledge, no such twins have been able to go on to lead independent, productive lives as a separated pair. After being asked to evaluate such a pair of infants for separation, our thoughts have undergone an evolution from the more commonly performed single-staged separation. In reviewing the available literature, it became clear that dividing the shared veins in a single stage was a likely source of brain injury. Furthermore, our experience in craniofacial surgery indicates that patients, especially infants and children, recover more easily when procedures are done expeditiously.

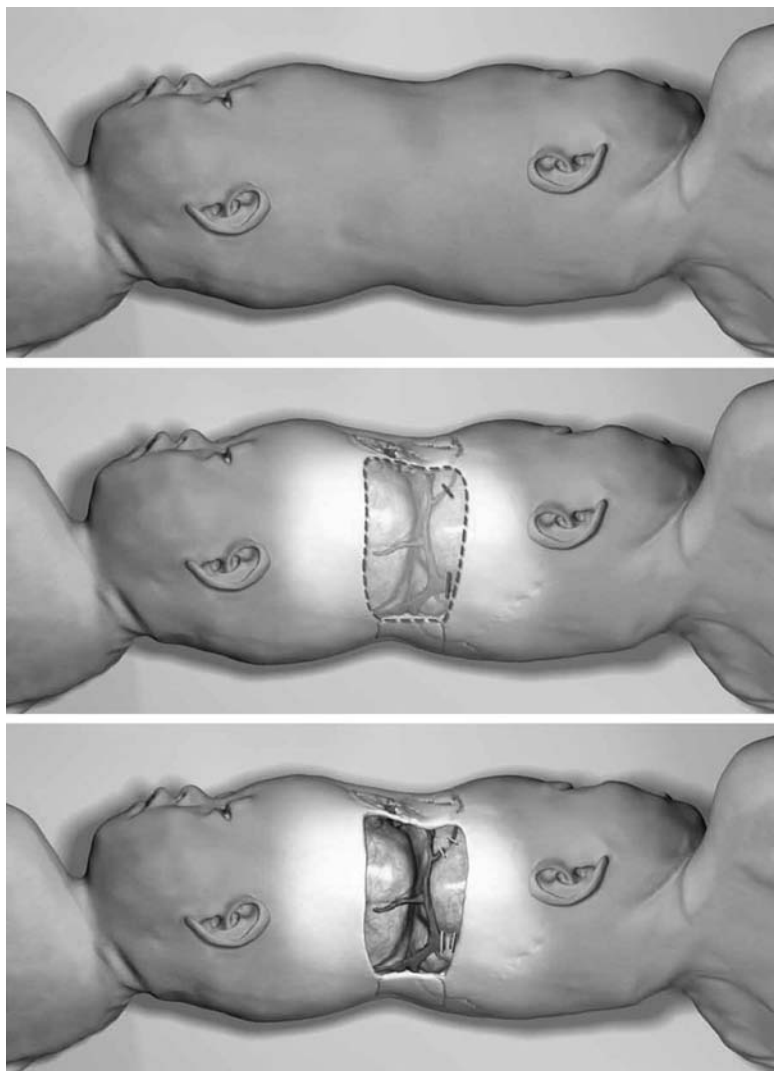


FIGURE 13 Note orientation and bridging veins that were ligated (clipped) and divided, leaving the circumferential venous sinus with twin A on left. *Source:* From Montefiore Medical Center/Medical Modeling, LLC, Golden, Colorado, U.S.A.

While the literature does include cases that have been separated in stages, these stages have been relatively close together and morbidity and mortality still appeared to be unchanged. We became convinced that a carefully planned multiple-staged separation could allow for the desired outcome if the patients were allowed appropriate recovery time between stages. After each stage we would not be able to predict the amount of time needed for recovery, but our team was committed to close observation and an open-ended schedule for the stages.

While the preservation of neurologic function became our prime goal, the difficulties of such a staged separation became clear. Tissue expansion has higher morbidity when performed at the same time as a craniotomy; vascular clips are needed on the edges of the dura, which then compromise a watertight closure leading to increased amounts of fluid around the tissue expanders. The limited area around the heads makes it difficult to place the expanders in a site where they are not exposed to this fluid and the resulting risk of infection and exposure. Because of our experience, we feel that expansion should be done as an isolated



FIGURE 14 Twin B postoperative day 14; he managed to remove his entire head dressing.

procedure prior to the final stage. Additional issues may arise as the soft tissue envelope is repeatedly elevated leading to some trauma, but this may be countered by a delay phenomenon as the flaps are elevated and returned to their original position, only to be used at a later stage.

The potential advantages of separation surgery in multiple stages include less exposure to general anesthesia, less bleeding, less intravenous fluids, less transfusion requirement, less cerebral edema, and less fatigue in the surgical team. The venous drainage between twins can be analogous to automobile traffic in New York City. During a single-staged separation, all of the bridging veins are ligated and divided during the course of surgery. This may be likened to closing off all the cross-streets in midtown at the same time, which would surely lead to "gridlock," and traffic would come to a stop. The circulatory equivalent in the brain is a frightful thought. However, if those same cross-streets were closed off gradually, drivers would learn to use alternate routes to get to their destination; existing roadways could be used and New York City would not need to build new roads. Surgically, we are encouraging the existing venous system in each brain to dilate gradually in order to handle the needed increase in flow.

In 2006, we consulted on another craniopagus in order to plan a similar staged separation in Europe. With the experience gained during our first staged separation of craniopagus, this second separation was successfully achieved in four major stages performed over a six-month period. The degree of involvement and specific anatomy was similar to the case reviewed. Again, both twins survived without neurological compromise (20).

For plastic surgeons, this concept and our experience with this separation, is particularly gratifying as we are constantly concerned about manipulating living tissue in order to maintain or improve function.



FIGURE 15 Twin A on postoperative day 14.



FIGURE 16 The separated twins on postoperative day 25; twin A on left. *Source:* From Montefiore Medical Center.

ACKNOWLEDGMENTS

All parties involved in the staged separation have donated their care. Philanthropic support was provided by:

- The Children's Hospital at Montefiore, Montefiore Medical Center (Bronx, New York, U.S.A.)
- Blythedale Children's Hospital (Valhalla, New York, U.S.A.)
- Medical Modeling, LLC (Golden, Colorado, U.S.A.)
- Voxel, Inc. (Provo, Utah, U.S.A.)
- Hill-Rom (Batesville, Indiana, U.S.A.)
- Inamed (Santa Barbara, California, U.S.A.)
- Children's Chance, Inc. (Waterbury, Connecticut, U.S.A.)
- Knightsbridge International (West Hills, California., U.S.A.)
- Philippine Airlines Foundation (Manila, Philippines)
- Hanger Prosthetics and Orthotics (Farmingdale, New York, U.S.A.)

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9 Orbital Dystopia

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DEFINITION

By definition, orbital dystopia is any type of abnormal displacement of the entire orbital cones and their contents that can occur in three different dimensional planes. Orbital dystopia in horizontal and vertical planes have been given the terminology orbital hypertelorism and vertical orbital dystopia (1,2). In defining orbital hypertelorism, Tessier (the father of craniofacial surgery) stressed the displacement of the entire orbital cones. The severity of the hypertelorism was based on the measurements of the intercantal distance and shape of frontal-orbital region (1). Following Tessier's principles, vertical orbital dystopia was defined as an abnormal displacement of the entire orbital cones in the vertical plane, not just displacement of one, two, or three orbital segments. Both orbital hypertelorism and vertical orbital dystopia initially referred to congenital conditions, but later were used to describe facial trauma, muscular torticollis, facial skeletal tumors, and iatrogenic and idiopathic causes (3). Nevertheless, it is extremely important to emphasize that the acute displacement of the orbital walls does not necessarily accompany the displacement of the orbital rims and should not be referred to as orbital hypertelorism or vertical orbital dystopia. In this line of thinking, facial trauma with a low impact force does not lead to orbital dystopia, mainly because there is not enough energy to provoke mobilization of the entire orbital cone. Therefore, it is our belief that, other than congenital and tumoral causes, only facial trauma with very high energy may cause displacement of the entire orbital cones.

HISTORY

The historical correction of orbital dystopia did not begin with Tessier. Nevertheless, his techniques in which the entire orbital cone must be osteotomized through craniofacial approach in order to adequately mobilize the orbits into a normal position introduced a new era in the craniofacial field (4). Before his principles, Converse and Smith attempted to accomplish the correction of orbital dystopia by mobilizing one segment of the orbit (5).

Tessier et al., subsequently followed by Schmid, were credited to have performed the first successful surgical correction of orbital hypertelorism, utilizing different approaches. Schmid assumed that the extracranial osteotomy of the orbital roof could not jeopardize the frontal lobe of the brain that lies just above the orbital roof in the anterior cranial fossa, and he successfully moved the orbital cones toward the midline (4,6). However, this method was not used by other surgeons.

Converse and collaborators performed a one-stage orbitotomy modification of Tessier's two-stage technique on four patients (7). Using one-stage technique, Tessier approached a large series of patients with orbital hypertelorism (1).

The first hemifacial rotation towards the midline through an intracranial route was first described by van der Muelen, who used the terminology "medial faciotomy" (8). This procedure allows movement of the entire facial halves through a combination of pterygomaxillary junction osteotomy and midline division of the bony palate, offering the advantage

of simultaneously approaching the malocclusion. Kawamoto noted that the majority of patients who underwent facial bipartition later require a Le Fort I osteotomy in order to accomplish a more normal occlusion. In skeletally mature patients with vertical dystopia, Kawamoto utilizes a facial bipartition procedure simultaneously with a hemi-Le Fort I (unilateral) in a one-staged procedure to obtain an optimal occlusion.

PREOPERATIVE PLANNING

Patients with the specific condition of orbital dystopia require a multidisciplinary evaluation that includes the following specialties; orthodontia, ophthalmology, psychology, neurosurgery, pediatric, and plastic surgery. The role of each professional in the care of patients with orbital dystopia should not be minimized.

Orthodontia

Prior to operative correction, patients with orbital dystopia have to be routinely seen by the orthodontist team. Frequently, the mal-positioning of the orbital cones have a direct impact on the maxilla, consequently leading to different types of malocclusion related to their malformation. Anterior open bite is frequently seen in patients with orbital hypertelorism mainly because the short ethmoid limits the caudal rotation of the maxillary alveolar ridge (9). In addition, a constricted maxillary arch with lateral cross-bite and high arched palate often exist. The orthodontic treatment also has an important role postoperatively to adjust the occlusion into a more normal position. Patients undergoing the traditional facial bipartition require an oral splint, especially if this procedure is performed in combination with a Le Fort I osteotomy as proposed by Dr. Kawamoto.

Ophthalmology

Patients undergoing correction of orbital dystopia require ophthalmologic evaluation to diagnose any type of visual acuity impairment due to amblyopia and/or extraocular dysfunction (10). Documentation of visual abnormalities is important to avoid correlation of any preoperative visual impairment to surgical procedure (this has medical repercussions).

Psychology

The central role of psychological preparation should not be underestimated. Families whose children undergo a major craniofacial procedure are usually threatened by the potential complications of the operation, and in many occasions this feeling is shown through anger, guilt, and fear. It is this behavior that has a tremendous implication in surgical expectations and outcomes. A team of psychologists working with technical expertise brings families a new perspective, helping them to better understand the whole process they are going to encounter.

Pediatrics

Patients with major craniofacial malformation usually have associated medical complications such as breathing problems and malnutrition, which directly interferes with their growth and health. In developing countries, this is an important issue since many patients with major craniofacial malformation (e.g., Apert and Crouzon syndromes, and facial clefts) present with anemia from chronic disease and malnutrition that warrant treatment prior to any operative procedures.

Plastic Surgery and Neurosurgery

Plastic Surgeons and neurosurgeons should work together in the planning of the orbital approach through an intracranial access. For this regard, the radiological documentation needs to be done preoperatively. Reformatted computed tomography (CT) and axial slices are indispensable for the preoperative planning. The CT scan also has a significant role in

detecting previous congenital brain malformation, which has a high impact in the patient recovery (11,12). Some studies have shown that a major craniofacial operation may worsen a previously existing CNS malformation (13). The neurosurgeon should be very attentive to this condition, and a contraindication of the major craniofacial procedure in cases of severe brain malformation should be considered.

Frontal cephalograms are used to measure the interorbital distance and to compare the data postoperatively. The final goal of the orbital hypertelorism correction is to achieve the optimal interorbital distance of 17 to 19 mm, but this interorbital distance may vary according to the age of the patient. The treatment of vertical orbital dystopia obeys the same principles as orbital hypertelorism and in most cases mobilizing the orbits in a vertical plane can be as challenging as mobilizing it horizontally.

The three-dimensional skull models provide surgeons with an additional tool for surgical planning, as well as a role in the education of young surgeons (Figs. 1 and 2).

Anatomical Considerations

In the preoperative planning, some anatomical considerations need to be discussed between plastic surgeons and neurosurgeons prior to the surgical procedure. Patients undergoing correction of orbital dystopia might present a combination of abnormal orbital cone displacement, and a series of correlated craniofacial malformations. The nasal cavity, ethmoid sinuses, and sphenoid wings are usually lengthened and the crista galli and cribriform plates are usually displaced inferiorly, particularly in patients with facial clefts. These complex craniofacial skeletal deformities may lead to a bone defect in the anterior cranial fossa. A herniation of brain and overlying meninges through the skull defect enhance the surgical challenge, and thus require meticulous preoperative planning. Patients with orbital hypertelorism and/or frontoethmoidal meningoencephaloceles may present an abnormal anatomical position of the



FIGURE 1 Dr. Kawamoto's commitment to teach craniofacial surgery to the future generation of surgeons. In this picture taken in 2006, he is using a three-dimensional skull model for facial bipartition planning at Sobrapar-Brazil.

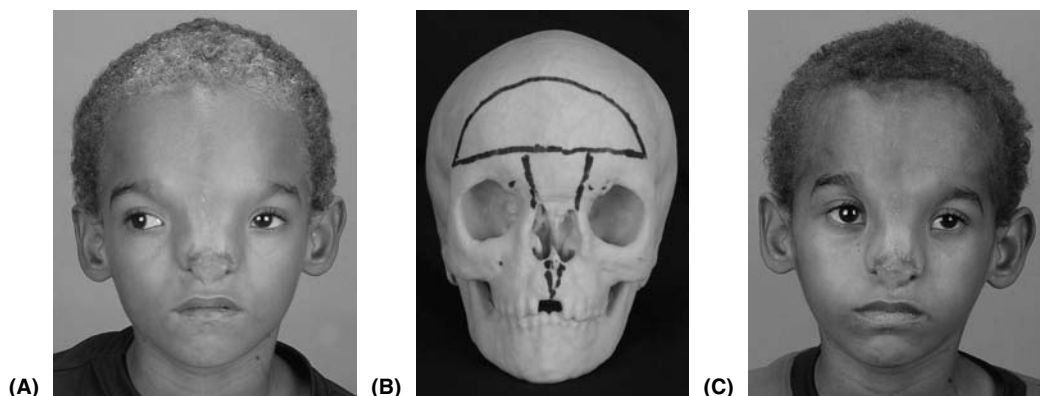


FIGURE 2 (A) Preoperative view of patient with a craniofacial cleft number 0 to 14. (B) Three-dimensional skull model marked for facial bipartition correction of hypertelorbitism. (C) Postoperative frontal view of same patient after facial bipartition and prior to nasal reconstruction.

cerebral venous sinus. These scenarios should be considered, because an iatrogenic injury to the venous sinus during the operation may be extremely difficult to control, even in experienced hands.

ORBITAL BOX OSTEOTOMY SURGICAL PROCEDURE

The following technique, originally described by Tessier, allows the mobilization of the entire orbital cones into a more normal position. This technique is routinely used for correction of vertical orbital dystopia, and is still used by some surgeons to correct orbital hypertelorism with normal occlusion (Figs. 3 and 4).

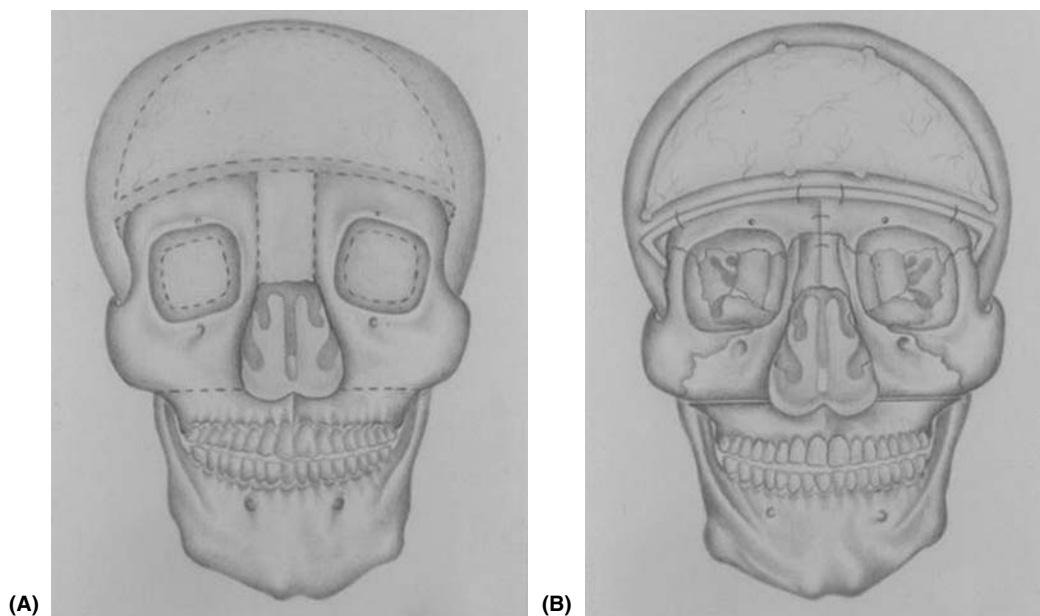


FIGURE 3 Correction of horizontal orbital dystopia (orbital hypertelorbitism) with orbital box osteotomy procedure. (A) Markings of osteotomies, including the frontal craniotomy and zygomatic, maxillary, and orbital osteotomies, and central bony resection are depicted. (B) Central translocation of orbits with fixation is seen.

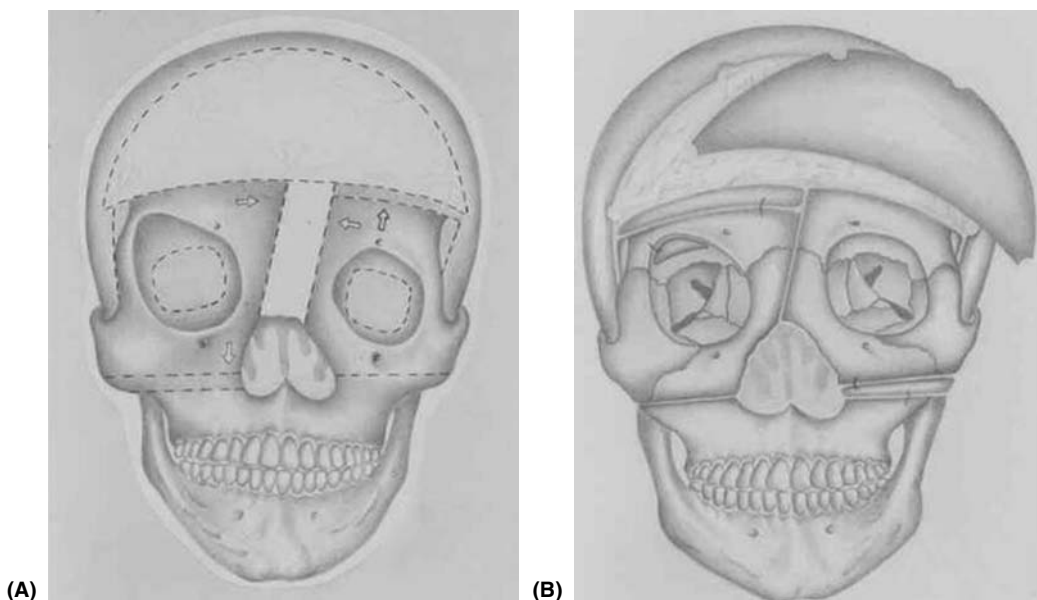


FIGURE 4 Correction of vertical orbital dystopia with an asymmetric orbital box osteotomy procedure. **(A)** Markings of osteotomies, including the frontal craniotomy, zygomatic, maxillary, and orbital osteotomies, and asymmetric central bony resection are depicted. **(B)** Central translocation of orbits with repositioning of full-thickness right zygomatic bone graft to right supraorbital bar and fixation is seen.

1. An oral ray endotracheal is used and suture-secured in the midline interdental.
2. After 0.25% marcaine and epinephrine injection, a coronal incision is performed.
3. Subperiosteal dissection is carried out over the supraorbital rims. The supraorbital nerve is carefully released from the supraorbital foramen using a 2 mm osteotomy. The orbital cavity and the nasal process of the frontal bone are carefully dissected to avoid injury to the optic nerve and to the nasolacrimal system, allowing orbital exposure for the subsequent osteotomy.
4. The temporalis muscle is left down and dissection is done along the superficial layer of the deep temporal fascia to the zygomatic body.
5. A gingivobuccal sulcus incision and dissection is used to complete the exposure.
6. A frontal craniotomy is performed by the neurosurgeon. Frontal bone flap is removed, allowing safe access to the orbital roof, cribriform plate, and crista galli.
7. A reciprocating saw through a temporalis muscle tunnel, is used to osteotomize the lateral orbital wall, and a malleable is used to protect the orbital content. Using an intracranial approach, osteotomies are continued through the frontal bandeau, then through the orbital roof.
8. The osteotomy is carried out along the orbital floor towards the medial orbital wall using a 3 mm osteotome. The orbital floor osteotomy needs to be as posterior as possible—up to 10 mm from the optic foramen—without jeopardizing the optic nerve. This is necessary to ensure proper translocation of the globes.
9. The zygomatic bones, including the zygomatic arch and maxilla, are osteotomized using a reciprocating saw, through a combination of a gingivobuccal sulcus incision and a coronal incision.
10. A V-shaped osteotomy is carried out in the midline, towards the crista galli. The frontal bone, ethmoid, and nasal septum are removed. At this point, any injury to the olfactory nerves may lead to a definite, undesirable impairment of olfaction. Kawamoto points out that this midline osteotomy does not necessarily need to be symmetrical, especially in



FIGURE 5 Preoperative photograph of a patient with orbital hypertelorism due to a number 1 to 13 Tessier craniofacial cleft (*left*). Postoperative photograph of the same patient (*middle*) and 23 years follow-up photograph. Besides the aging phenomenon, a slight relapse of the bony orbits into a previous position is seen (*right*). (Surgery performed by Dr. Raposo do Amaral in Brazil 26 years ago.)

patients with craniofrontonasal dysplasia and orbital dystopia in all three-dimensional planes. In these cases, an asymmetric V-shaped wedge excision is necessary.

11. The orbits are translocated to the midline, and rigid skeletal fixation is performed with titanium plates and screws.
12. Bilateral medial canthopexy is performed when necessary, using transnasal wire fixation. In addition, transnasal wires for medical canthal soft tissue bolsters are helpful to correct soft-tissue fullness and telecanthus (14,15).
13. A cranial bone graft to the nasal dorsum for improved nasal projection and lengthening is also sometimes needed.
14. The coronal incision is closed in two-layer fashion, and the oral mucosa is closed with running-locking chromic (Figs. 5 and 6).



FIGURE 6 Preoperative photograph of a patient with vertical orbital dystopia due to a severe unilateral coronal synostosis in combination with a mild muscular torticollis (*left*). An orbital translocation into a more superior position was achieved by a craniofacial approach. Eight (*middle*) and twelve years (*right*) follow-up photographs of the same patient. (Surgery performed by Dr. Raposo do Amaral.)

FACIAL BIPARTITION SURGICAL PROCEDURE

The facial bipartition procedure performed in combination with medial canthopexy allows for the hemifacial rotation towards the midline and correction of the position of the lateral canthi. It is indicated for orbital hypertelorism patients with an inverted V palate shape and malocclusion. This procedure is preferred by the majority of craniofacial surgeons to correct orbital hypertelorism. In skeletally mature patients, Dr. Kawamoto has used facial bipartition in combination with a Le Fort I osteotomy to maintain a good occlusion. Combining procedures allows the mobilization of the upper jaw into a more normal occlusion based on the preoperative orthodontic planning. This requires a definitive oral splint that should be used for six weeks. The facial bipartition procedure may also be used simultaneously with an acute monobloc advancement of a monobloc distraction when midface hypoplasia exists (in patients with craniofacial dysototsis syndromes). With regard to the operative technique, the intracranial exposure and orbital osteotomies are similar to the facial bipartition operative technique compared to the orbital box osteotomy procedure as detailed below (Fig. 7).

1. A zig-zag coronal incision is made, the sub-periosteal dissection is used, and the coronal flap is turned down. The temporalis muscle is left down (to avoid temporal hollowing postoperatively) by dissecting just above the superficial layer of the deep temporal fascia (a plane just below the frontal branch of the facial nerve).
2. A small subperiosteal tunnel is made under the temporalis muscle for the lateral wall osteotomy with a reciprocating saw. Next, all osteotomy sites are exposed with subperiosteal dissection around the circumferential orbit, caudad to the nasofrontal suture and to the zygomatic body. Intraoral exposure with small bilateral gingivobuccal sulcus incisions is used for the pterygomaxillary osteotomies.
3. Craniotomy of the forehead bone flap is performed and, with retraction of the dura, the orbital roofs are exposed.
4. Osteotomies are performed bilaterally in the anterior zygomatic arch, lateral orbital wall, orbital roof, medial orbital wall, orbital floor, pterygomaxillary buttresses, and septum (after the mid-face down-fracture is started). Specifically, the lateral orbital wall osteotomy

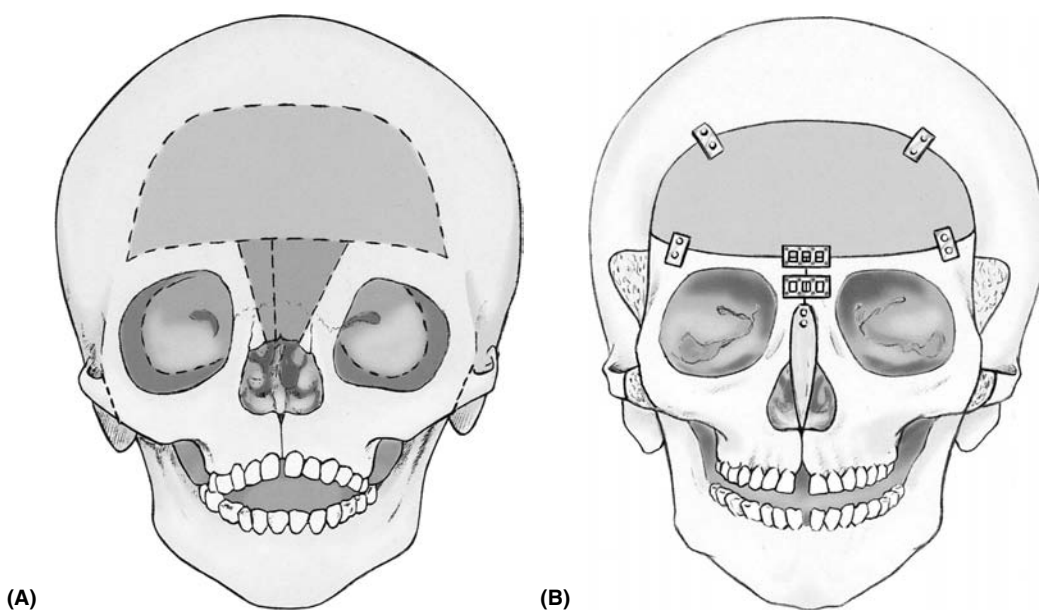


FIGURE 7 (A) Frontal view of osteotomy lines, including craniotomy, midline asymmetric "V" wedge excision of frontonasomethmoidal bone, and midface buttresses (zygomatic arch, circumferential orbital walls, pterygomaxillary). (B) Frontal view of fixation with midline "keystone" box fixation.

is made with a reciprocating saw passed underneath the temporalis muscle and directed toward the inferior orbital fissure. The lateral orbital wall osteotomy is carried up to the orbital roof through the sphenoid bone, then along the roof and across cranial base anterior to the cribriform plate at the midline. A contralateral and symmetric osteotomy is made and connected to this midline osteotomy.

5. Next, a small (4 mm) osteotome is used for the orbital floor 1 to 2 cm posterior to the orbital rim and behind the lacrimal crest. The same osteotome is used for the medial wall osteotomy behind the medial canthus to complete the "doughnut osteotomy" around the globes.
6. The Kawamoto osteotome is used to make the ptergomaxillary osteotomies bilaterally.
7. Just prior to the monobloc down-fracture the intradacyron distance is measured with an orbital caliper. A V excision is marked between the orbits in order to leave 16 to 20 mm of bone between the medial canthi region. This V includes the mid-portion of the supra-orbital rim, the radix, and the central portion of the nasal bone. The V is planned so there is a rotation point centered just above the nasal spine. For asymmetric hypertelorbitism, an asymmetric V excision is planned.
8. Next, intraorally a midline 2 cm gingivobuccal sulcus incision is made and subperiosteal dissection is performed to split the palate.
9. Next, the monobloc down-fracture is performed with the Rowe disimpaction forceps. (For a high arched palate, an acrylic splint is used to avoid fracturing or splitting the palate).
10. During the down-fracture the double-pronged septal osteotome is introduced within the nasofrontal separation and is directed toward the hard/soft palate interface to cut the perpendicular plate.
11. For full mobilization, the Rowe forceps are used to stretch the midface from side-to-side while the Kawamoto osteotome provides anterior traction in the ptergomaxillary osteotomy.
12. After the monobloc downfracture, the previously marked midline V is excised with the reciprocating saw. (It is important to remove bone above and behind the medial canthus to allow the greatest correction; however, a stable medial canthal attachment to the bone is crucial.) The V is then cut and the bone is removed.
13. Through the previously dissected midline gingivobuccal sulcus incision the hard palate is split with a reciprocating saw. A fine osteotome is used to complete the midline osteotomy between the central incisors.
14. The bipartition halves are then brought together by rotating them to the midline. A convexity to the supraorbital region is created. (Both horizontal and vertical correction may be done at this time, depending if symmetrical or asymmetrical hypertelorbitism exists.)
15. Midline fixation is initially made with a 24-gauge wire through drill holes made into the stable bone lateral to the excised V. The intradacyron distance reduction is confirmed with measurement with an orbital caliper. Final rigid fixation is made with a "box" titanium plate.
16. For monobloc distraction using internal devices, the proper convexity (Roman arch curvature) of the supraorbital region provides the necessary stability for advancement (16).
17. When performing a facial bipartition in skeletally immature patients with asymmetrical hypertelorbitism, maintaining a proper occlusal plane is not critical; however, for the mature patient, a hemi-Le Fort I osteotomy is necessary at the end of the procedure to achieve a good occlusion).
18. A pericranial flap (10 cm × 4 cm) is sutured into the midline defect to separate the nasal sinus cavities from the epidural dead space. Fibrin glue is then used over the flap.
19. The frontal craniotomy is then reshaped as necessary and attached to the advanced frontal-orbital rim using 1.0 microscrews and plates.
20. Soft tissue refinements are performed. Medial canthopexy positioned high and posterior is performed at times. Transnasal medial canthal soft tissue wires (26 gauge) often are placed in a deep and superior position of the medial orbit and bolstered externally with xeroform gauze.
21. Next, the K-stitch is performed (15). Redundant subcutaneous soft tissue is excised in the central area. Then, with the coronal flap repositioned, an assessment is made to the extent

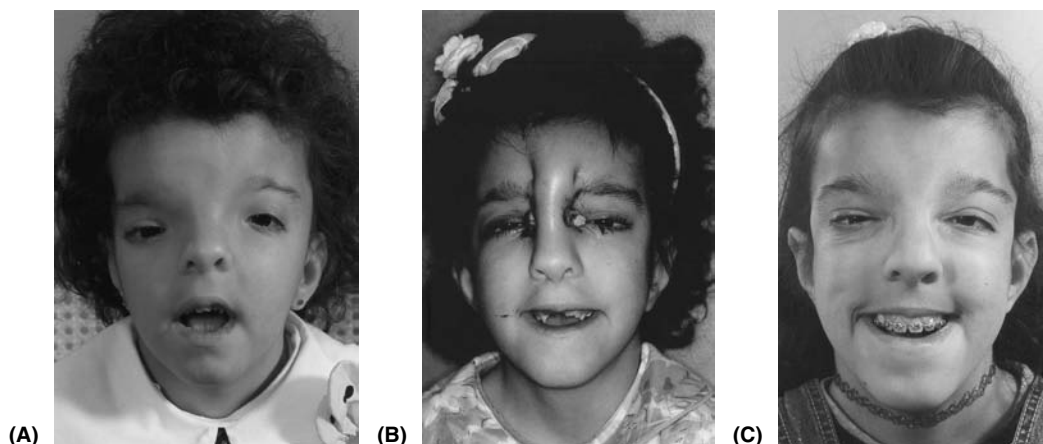


FIGURE 8 A nine-year-old female with asymmetric hypertelorism. (A) Preoperative frontal view. (B) One week postoperative photo with soft tissue medial canthi bolster and “K-stitch” in place. (C) Three-year follow-up frontal view demonstrates reduced intercanthal and interbrow distances with minimal scarring following the “K-stitch” procedure.

of glabellar skin excess between the eyebrows. A horizontal mattress suture with 3-0 vicryl is placed in the hair-bearing skin of the medial eyebrow. The suture is tightened until the eyebrow position is just medial to the medial canthus. This suture causes the excess soft tissue to bunch-up in the center of the forehead. Over a 6-week period, this excess will subside.

22. The coronal and givibuccal sulcus incisions are closed, and drains are usually used (Fig. 8).

TIMING AND FOLLOW UP

Appropriate timing for either vertical orbital dystopia or orbital hypertelorism is still a controversial subject in craniofacial literature. Mulliken et al. highlighted that orbital hypertelorism grade III has the potential to relapse over 5 mm throughout the years, attributing neither to age, diagnosis, nor orbital anatomy as a cause for this undesirable phenomenon. They suggested to postpone the mild orbital hypertelorism cases until late adolescence in order to assure normal facial growth (17). On the other hand, McCarthy et al. advocated that the orbital hypertelorism could be safely performed before age 5.3 years (average age 3.9 years), demonstrating desirable results and little orbital skeletal relapse throughout the subsequent years. He reported a final goal of 15.5 mm of interorbital distance in a population of children with less than five years of age. The discrepancy of skeletal orbital relapse measurement between the authors was attributed to a different definition of orbital skeletal relapse (18). It is our preference to operate on patients with either orbital hypertelorism or vertical orbital dystopia when they are between 8 and 12 years of age. Nevertheless some degree of orbital translocation or relapse may occur in long-term follow up.

Tessier stressed that correcting the associated soft tissue malformation in patients with orbital hypertelorism can be even more challenging than the approximation of the bony orbits. Kawamoto modified the hypertelorism correction with the soft-tissue retirement of the K-stitch. This method obviates the need for a midline forehead-scar. The combination of orbital hypertelorism and vertical orbital dystopia, commonly seen in patients with a craniofrontonasal dysplasia and unilateral coronal synostosis leads to a challenging three-dimensional deformity. Careful preoperative planning is required and if it is also associated with a muscular torticollis, a cranial skull base rotation can be observed and total post-operative facial symmetry is almost impossible to achieve.

ACKNOWLEDGMENT

We express our deep gratitude to Dr. Kawamoto. He has been an envoy of hope for the Sobrapar institute in Brazil.

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10 Cleft Lip and Palate

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BACKGROUND

Clefts of the lip and palate have presumably existed throughout human history. Descriptions of surgical repair of cleft lip (CL) have been found dating to 3rd or 4th century China (1). The second most common (about 1 in 600 live births) congenital deformity, after club foot, these localized clefts produce multiple regional clinical challenges: they affect appearance, speech, and deglutition, as well as increase the frequency of ear infections and cause secondary problems of social stigmatization and decreased self-esteem.

Isolated cleft palate (CP) occurs with a genetic pattern distinct from that of CL (with or without CP—CL/P). The incidence is 0.45% of live births, with a male:female ratio of 0.73 (in Caucasians) (2), while the occurrence of CL/P varies among genetic groups—0.3 per 1000 in blacks, 1 per 1000 in Caucasians, 1.4 per 1000 in Chinese, 2.1 per 1000 in Japanese, and 3.6 per 1000 in Amerindians (3–5). The M:F ratio of CL/P is 2:1 and of CL is 1.55:1 (in Caucasians) (2). The incidence of CL is probably affected by nutrition: folic acid supplementation has been shown in some studies to be beneficial in decreasing the recurrence risk (6). It has been suggested that the variable expressivity of palatal clefting in the presence of lip cleft is because a cleft of the lip alters tongue-palate relationships, such that the tongue may secondarily obstruct palatal closure during facial development (7). Conversely, Gorlin has suggested that it arises instead from a reduction in size of both the labiomaxillary prominences and the palatine process of the maxillary prominences (3).

Most clefts occur in isolation and without known etiology. In a study of California clefts identified among 2.5 million births, 62% were sporadic (2). Debate continues as to the relative contribution to the etiology of clefts of genetic, environmental, and developmental factors. The multifactorial threshold model appears to have lost some currency, as it has failed to live up to the facts of published data, particularly for CP (3). Consistent, however, with the suggestion that a familial genetic predisposition to clefts underlies some “sporadic” cases are reports of consistent mildly abnormal embryonic facial morphology in mice genetically inclined to clefts of the primary palate (7). A statistical analysis of recurrence patterns in CL/P in families containing varied numbers of clefts suggests that the interaction of perhaps three to six genes may be responsible for the cases studied (8). *MSX1* and *TGF β3* allelic variants are among the most suspect genes, especially when present in mothers who use tobacco or alcohol (3). While we await results of the current frenzy of genomic investigation, the near-term elucidation of the genetic sources of the defects, it is far from clear that such information will yield a satisfyingly simple explanation.

There are over 350 known syndromes associated with clefts (3), of which half are of monogenic origin (7). A significant majority of the remaining syndromic cases are due to Pierre Robin sequence (25% of syndromic clefts) and velocardiofacial (VCF) syndrome (15%) (9). The Pierre Robin sequence is associated with the majority of cases of Stickler syndrome.

Care of affected children by specialized “cleft teams,” a concept originated by Tessier and disseminated around the world by his disciples, has been accepted by the medical profession, if not always by third-party payers, as the standard of care. The team consists

of, at a minimum, a plastic surgeon, an orthodontist, and a speech pathologist. More generally, a team also includes the following: pediatrician, otolaryngologist, oral surgeon, geneticist, genetic counselor, registered nurse, audiologist, pediatric dentist, prosthodontist, ophthalmologist, social worker, radiologist, pediatric anesthesiologist, and sometimes psychologist and anthropologist. The coordinated evaluation and treatment that a team can provide results in the most efficient provision of complete care to the cleft patient and family. With team care, interventions can be best sequenced and, as appropriate, combined into fewer operative stages, to optimize quality and minimize cost.

CLEFT DEFICIENCIES

The anatomic deformity of a cleft can be as minimal as a bifid uvula or a groove in the vermillion in line with the peak of Cupid's bow. Ultrasound studies of families of cleft probands (7) have shown even smaller defects, with subclinical orbicularis oris muscle deficiencies. At the opposite extreme "complete" clefts can include a superimposed set of gaps in skin, lip mucosa, subcutaneous and submucosal tissue, orbicularis oris muscle, gingiva, alveolar bone and teeth, palatal roof and nasal floor mucoperiosteum, hard palatal bone, and soft palatal mucosa and muscle. There is additionally an associated nasal deformity, including deficient bony support of the nasal base and deformity of the lower lateral cartilage. Thus, the entire caudal hemi-nose is affected, bilaterally in a bilateral cleft. A complete palatal cleft therefore means the absence of side-to-side bony continuity of the face from the mouth up to the level of the nasal bones, with resultant instability and perhaps deformity of the entire midface, almost up to the level of the inferior orbits.

BRIEF HISTORY OF SURGICAL TREATMENT OF CLEFT LIP

The early history of surgical repairs of unilateral CL has been well described by Still and Georgiade (10) and by Rogers (1), with clear diagrammatic sketches by the former. Repairs in the West date back at least to 10th century Britain and to Yperman's 14th century description and in the East to 3rd century China (1). Nineteenth-century and earlier techniques of CL closure by V excision or by vertical closure of horizontal incisions were frequently followed by excessive vertical scar contraction and were supplanted by the "straight-line" repairs of von Graefe, Rose, and Thompson. These curve the excision lines to lengthen the vertical line of closure (see sketches in Ref. 10). Malgaigne in 1843 began the use of local flaps that led to the Z-like techniques of Koenig and Hagedorn (10). The Z functions to break up the straight line of closure and thus diminish the tendency towards vertical scar shortening. It also causes less horizontal tension in the lip. The most widely used repairs of the late 20th century were more advanced applications of the Z concept, primarily those of Tennison (modified by Randall and by Bardach) (11) and Millard (12), with some surgeons still preferring the techniques of LeMesurier or Skoog. The Tennison repair before Randall's modification has been criticized for sometimes producing lips that are too tall. Millard's rotation-advancement repair, with its placement of the scar along at least the caudal portion of the desired course of the philtral column, has gained widespread popularity.

Cutting has documented the ability to produce lips of normal height and nearly symmetrical width following a modification of a rotation-advancement repair by using the c flap to fill the entire rotation defect and by locating the caudal end of the advancement flap far enough laterally to make the distance from columella to vermillion match the height of the contralateral lip at the corresponding peak of Cupid's bow (13). This report notwithstanding, the persisting presence, including in published works, of a number of vertically short lips testifies to the variability of technique and judgment with surgeons and perhaps to the innate limitations of the rotation-advancement technique (14).

In recent years, several surgeons (13,15,16) have reported success with rotation-advancement-like repairs that move the post-rotation defect of the medial lip element superiorly all the way up to or even into the columellar. This positions the lip scar line along the desired philtral column location for the entire lip height. Salomonson (15) uses a Z-plasty to fill the gap in the

columella. Salyer (16) reports the “rare” need for additional lip height at the time of lip repair, achieved by placement of a small triangular flap at the level of the white roll. Cutting (13) reports a 3% incidence of short lips with his approach. Measurement of lip height at the time of surgical repair shows clearly that the cleft side is always shorter than the normal side. Therefore, the success of these authors suggests that the lip will stretch under the influence of neighboring tissues.

Good results following lip and palate repair have been reported by many surgeons using a variety of surgical techniques. The presence of additional reports of mediocre results suggests that an individual surgeon’s training, technical abilities, and experience have a notable effect on ultimate outcomes. In consequence, individual reports of good or bad results, even in prospective studies, are not in themselves reason to use or to avoid a particular technique. This variation in outcome following use of ostensibly the same operative technique, in combination with the requisite years-long follow-up of treatment of a significant number of patients with a particular treatment protocol, accounts for our persisting lack of certainty of the best treatment protocols for cleft patients. A confounding element is the variability between the clefts of different patients, which arguably require individualized surgical treatment rather than an identical protocol for all.

Each surgeon should use the procedures that provide him/her the best clinical results. The goals of CL repair remain constant: symmetrical vermilion of normal height, with a well-shaped Cupid’s bow and a full (convex) tubercle; symmetrical white (supra-vermilion) part of the upper lip, including symmetrical vertical position of the nasal alae; a distinct and continuous white roll; a concave philtral profile (i.e., mid-sagittal) as part of a symmetrical and concave philtrum of normal width and with pleasantly-contoured (negatively curved) columns; negatively curved shape of the caudal portion of the lateral segments of the white portion of the lip (concave in sagittal plane, convex in horizontal plane); maximum lip projection in all sagittal planes at the vermilion–cutaneous (V–C) junction; imperceptible scars; normal appearance (symmetry, no dimpling) during motion.

UNILATERAL CLEFT LIP REPAIR

For the incomplete cleft the Millard rotation–advancement repair is usually ideal. For the complete cleft, however, the primary author has found the Davies (and the modification by Lewis) repair most useful (17). It attacks the need for a Z head-on by putting it near the vertical center of the lip, making it as large as necessary to produce a lip of fully normal height. By this generous form of tissue arrangement, transverse tightness of the lip is minimized. The primary author finds this repair the one most likely to consistently give a normal lip shape, albeit one that has a scar on it, with the least likelihood of eventual shortening.

Marking

Once general anesthesia has been induced, landmarks are marked with ink and then tattooed (Fig. 1). I find this easiest to do with a 25 gauge 5/8 inch needle attached to a 1 cc syringe containing only 0.1 to 0.2 ml of ink.

- Local lowpoint (presumed midline) of V–C junction on the prolabium
- V–C junction at peak of Cupid’s bow on the normal side
- On the medial side of cleft, the corresponding location of the symmetrically-positioned peak of Cupid’s bow, located by inspection and verified by measurement from the midline mark
- On the lateral lip element, the V–C junction at the termination of the white roll, or the nearest location where the vermilion is full-height
- Both right and left points of termination of the columella at the upper lip
- Points of termination of both right and left nasal alae at the upper lip, which point is accentuated by passive lifting of the subjacent lip.

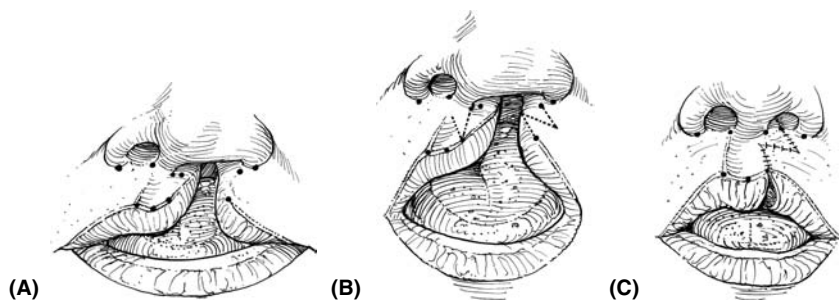


FIGURE 1 Davies-type repair of unilateral cleft lip. **(A)** The lip landmarks are tattooed: the V–C junctions of the prolabial midline, the peaks of Cupid's bow, and, on the cleft side, the most medial location of full-height vermillion; the junctions of lip with bilateral columellar margins and alae. Also marked are points several mm towards the cleft from both the columellar base and the alar base, such that the sum of the two medial displacements equals the width of the nostril sill on the normal side. Also, these two dots are to be marked slightly caudally, as the columella and cleft-side ala typically slope caudally towards the cleft. **(B)** On both sides of the cleft two limbs of an equilateral triangle are drawn, in preparation for a Z-plasty. The superior ends of the limbs adjacent to the cleft connect to the non-anatomic dots noted above. There is an error in this drawing in the marking of the lateral Z limb on the medial side of the cleft: at its superior end it should instead connect to the dot that lies closer to the cleft. **(C)** The Zs are cut full-thickness through the lip. The lateral lip element has been vertically shortened to make its height match the length of the other Z limbs by full-thickness excision of a narrow triangle of lip. The Z flaps are transposed and the lip closed in layers.

The height of the normal side of the lip is measured with caliper from the tattooed Cupid's bow peak vertically to the unmarked line that connects along the nostril sill the two dots at nasal ala and columella. This is the target height for the cleft side. To construct the Z, two limbs of an equilateral triangle are drawn on both the medial and lateral lip elements, with one limb in each case placed roughly vertically along the cleft margin. One must plan the horizontal distance along the cleft-side sill from ala to columella to match that of the normal side of the lip, and this is determined by the position of the superior ends of the two vertical limbs, as will be described.

On the medial element, the V junction of limbs is caudal, at (or above—v.i.) the selected peak of Cupid's bow. One limb goes nearly vertically, and the other limb, of equal length, travels in a straight line superomedially from the same point. The distance between the superior ends of the two limbs should be a limb-length, to ensure 60° angles (18). If the lip height here is taller than needed for the Z limb, the V junction is positioned instead more superiorly, and the lip intervening between the Cupid's bow peak and the V junction is excised as a full-thickness narrow triangle. In the generic case, this caudal junction of the two limbs may therefore be remote from the desired location of the Cupid's bow, until the narrow triangle is excised. It is in fact uncommon for the medial lip element to be tall enough to require this excision.

On the lateral element, the V junction is superior, adjacent to the nostril sill. The two limbs pass down from the point on the junction of nostril sill and lip that is at the medial edge of that lateral segment. One limb passes along the cleft margin and the other inferolaterally at 60°. Analogous to the design on the medial lip, if the lip height here is taller than needed for the Z limb, the V junction is positioned instead more caudally, and the lip intervening between the nostril sill mark and the V junction is excised as a full-thickness narrow triangle. Again, the limb ends are a limb-length apart.

The length of all limbs is $0.59 \times$ the height of the normal side of the lip. Empirically, I generally add 1 mm to the contralateral (normal) lip height before calculating Z limb length (19).

Thus, if the height of either the medial or the lateral lip element exceeds the required limb length, this segment is reduced in height via excision of a narrow full-thickness triangle of tissue adjacent to the oblique limb, i.e., at the caudal end of the medial element or at the superior end of the lateral element, displacing the V junction accordingly.

Alternatively, if the lip height is shorter than that required for the Z, a lip adhesion is done, and the patient is returned to the operating room (OR) for definitive repair when this matures and softens.

Triangular-tipped stainless steel templates of various sizes designed to facilitate the Davies repair are readily available from instrument vendors, to mark a V with equal limbs at a 60° angle, but it is easy enough to mark them free-hand.

The senior author has found that the advancement flap has fulfilled a wide variety of clinical needs for reconstruction of both complete and incomplete clefts. Surgical descriptions are conveniently found in a large selection of plastic surgery resources.

Repair

Lip repair is done at any time after age three months (adjusted for prematurity), as long as the child is healthy and can medically withstand general anesthesia.

The repair is performed with loupe magnification. After being tattooed and marked, the lip is infiltrated with a suitable vasoconstrictor, typically xylocaine 0.5% with epinephrine 1:200,000. Time is allowed to elapse for vasoconstriction. The limbs are incised with a #15 or #67 scalpel and cut full-thickness through the lip with a #11 scalpel. Appropriate care should be taken to avoid narrowing the deeper tissue. The vermilion may (or may not) be left in excess at this time, to be trimmed during final suturing. The muscle is not dissected out from the flap, but the lip flaps are rather to be left as intact segments. One may reshape the nasal tip at this time if desired. Specifically, the alar cartilages may be dissected and sutured via the columella and alar base through the existing wounds, sometimes with the addition of a rim incision. The Z flaps are then transposed and the lip closed in layers with absorbable suture. It is important to snug the tips of the flaps, especially the muscle layer, into the corners in order to obtain all possible height from the Zs. I typically reinforce the deep dermis with five to zero Maxon and close the lip with six to zero fast-absorbing plain gut. Some surgeons prefer to use five to zero Vicryl and reinforce the skin with Steri-Strips.

After completion of the closure, the central limb of the Z lies horizontally across the lip. This limb is the one most likely to spread during healing and therefore justifies placement of three sutures in the muscle layer.

Postoperative Care

Elbow splints are placed before the patient leaves the OR. Parents are instructed to peek at the covered skin daily to look for rash but to otherwise leave the splints on continuously for two weeks (20). Skin irritation can be prevented or managed by having the parents apply under the splints a wrap of cotton T-shirt material. Non-syndromic children are almost always discharged home the same day. Children with syndromes are kept overnight. Infants are placed in a car seat in the Recovery Room and for several days after surgery to minimize swelling, bleeding, and pain. They may return to bottle or breast immediately.

BILATERAL CLEFT LIP REPAIR

The greater tissue deficiency of a bilateral cleft makes the task more challenging than with a unilateral cleft, but the goals of treatment as stated above remain the same. As with unilateral clefts, the extent of clefting can vary from a mere groove in the vermilion to a wide complete cleft of all layers of lip and palate.

Treating the associated nasal deformity of the bilateral cleft is arguably a more significant topic than repair of the lip itself. This is truer now than at any other time in the last half century, given the large amount of ferment in the literature. Evidence is accumulating to suggest that results of treatment of the bilateral CL nasal deformity may be forever limited if it is not done at the time of primary lip repair. A detailed discussion is beyond the scope of this chapter. Briefly, McComb popularized the concept of reshaping the alar cartilages with sutures after extensive separation from the sandwiching nasal tissues (21). The increasing availability of sophisticated pre-operative orthodontia can allow pre-surgical stretching of the philtrum prior to cartilage repositioning (22). Mulliken suggests that the nose already contains sufficient tissue to allow creation of a columella without prolabial stretching, by appropriate freeing and suturing of the alar cartilages (23). This approach appears to be finding more

adherents and may represent the path that will define 21st century treatment of the bilateral CL nasal deformity (24,25).

Some of the more challenging patients are those with asymmetrical clefts. The question arises as to whether to treat the minor side with a full-height straight-line repair or to do a less extensive repair. Plastic surgeons are sometimes tempted to do a rotation-advancement or other unilateral repair on the major side. Usually, the best appearance comes from using a symmetrical straight-line repair, although even here asymmetry tends to remain because of the difference in tissue volume on the two sides.

Preoperative Orthopedics

Because of the typical presence of premaxillary protrusion and the consequent increased distance between the lip segments, it has become more and more common to use pre-operative mechanical techniques to posteriorly reposition the premaxilla. Pinned palatal appliances have been pioneered by Georgiade and Latham (26) and have evolved into the naso-alveolar molding techniques described by Grayson (27). Socio-economic pressures on our patients and on our orthodontists have thus far precluded the use of these devices in our patients. We have often used several pre-operative weeks of simple taping techniques from cheek to cheek across the prolabium with the well-tolerated Micropore tape (3M, St. Paul, Minnesota, U.S.A.). To protect the cheek skin from the repeated removal of cell layers, we place a base layer of tape on the cheeks and then tape to the tape, replacing the base layer only when it wears away or comes off.

Marking

Once general anesthesia has been induced, landmarks are marked with ink and then tattooed:

- Local lowpoint (presumed midline) of V-C junction on the prolabium
- V-C junction at desired location of the peaks of Cupid's bow on the prolabium, 3 mm from the midline
- On the lateral lip elements, the V-C junction at the termination of the white roll, or the nearest location where the vermilion is full-height
- Right and left points of termination of the columella at the upper lip
- Points of termination of both right and left nasal alae at the upper lip, which point is accentuated by passive lifting of the subjacent lip.

A superiorly-based philtral flap is outlined on the prolabial midline. The flap should have slightly concave lateral borders, as there is a tendency for the healed flaps to widen over time. Should it appear that there is likely to be substantial tightness of closure, which is of course more common without pre-operative orthopedics, the prolabium should instead be left full-width, with plans to narrow it at a later time. In this latter situation the prolabial mucosa and even vermilion can be turned laterally and sewn to the mucosa of the lateral lip elements, and the orbicularis oris musculature should then be sutured only to the subcutaneous lateral edges of the prolabium rather than across the midline with the preferred muscle-to-muscle closure. Muscle connection across the midline will then be done secondarily, at the time of narrowing of the neo-philtrum.

The caudal edge of the prolabial flap is marked as a bilobed scallop, with the intent of using as the white roll the scar at its junction with the subjacent vermilion flaps from the lateral lip elements. Laterally-based vermilion flaps are designed on the lateral lip elements, based at the end of the white roll and extending medially 1/2 to 1 mm longer than half the prolabial flap tip width, thus usually 4 mm. It is alternatively appropriate to leave these long and trim them before their final closure together in the midline.

Bilateral flaps are marked on the lateral lip elements as a horizontal line at the level of the alar bases, connecting to an oblique line along the V-C junction.

Repair

After tattooing, the lip is infiltrated with xylocaine with epinephrine for its vasoconstrictive effect. The lateral segment vermilion flaps are raised with the scalpel of choice. L flaps from the vermilion remaining more medially are usually discarded.

The superiorly-based philtral flap is raised at the level of periosteum up to the junction with columella. The remaining white skin and prolabial vermilion on either side of the philtral donor site are excised and discarded; currently, most surgeons do not use forked flaps for columellar lengthening. The prolabial mucosa is tailored across the premaxillary labial surface to help form a labial sulcus. The orbicularis oris muscle is dissected from the nose and from the skin and mucosal surfaces of the lateral lip elements sufficiently lateral to allow it to be brought to the midline. However, excessive skeletonization may lead to devascularization and should be avoided when tissues are deficient, in favor of securing it to the lateral margin of the unviolated prolabium. The mucosa from the lateral segments is sutured together in the midline. The muscle is repaired together in the midline with absorbable suture and then secured with a permanent suture at its superior margin to the periosteum at the anterior nasal spine.

The neo-philtrum is now closed to the surrounding vermilion flaps and lip skin of the lateral segments. Care is taken to precisely align the V-C junction and the wet line. Post-operative care is as described for the unilateral CL.

BRIEF HISTORY OF CLEFT PALATE REPAIR

The earliest reported repair is that of LeMonnier in the 1760s (28), who placed sutures and then cut the cleft edges. This was followed by suppuration and then healing. Credit, however, usually is given to von Graefe (1816) and Roux (1819) (29). Von Langenbeck introduced the use of mucoperiosteal flaps to close the hard palate (30). Currently, the most widely used repairs of the hard palate are Von Langenbeck's, the Veau-Wardill-Kilner (VWK), and the two-flap repair as described by Bardach (31). The "push-back" concept of the VWK and the original Dorrance repair, intended to lengthen the soft palate, has not been convincingly shown to actually yield such lengthening (32,33). Soft palatal repair is commonly done by either Furlow's double Z-plasty (34,35) or (other form of) intra-velar veloplasty (IVV).

The IVV, originally proposed by Kriens (36), was adopted by Bardach (31). Cutting, who learned it from Bardach, has reported excellent results with an extensive dissection of the muscle (37). Less aggressive IVV was shown no better for speech than simple suturing together of undissected muscle in a much-discussed prospective study by Marsh (38).

Persisting areas of critical concern to the CP surgeon have for some years been outcomes of speech and of facial growth. Most reports suggest that facial growth of an individual with unoperated CL and palate is normal, although some disagreement persists (39). Surgery on the hard palate probably causes some limitation of anterior growth of the maxillae. When hard palate closure is delayed until six years of age, maxillary growth is believed to approximate normal.

Speech may develop normally when the palate is closed by age 12 months. Delays beyond 18 months can lead to abnormal speech patterns. The chance of development of normal speech becomes progressively less likely as establishment of reasonable anatomic normality of velopharyngeal structures is further delayed. It remains in doubt whether normal speech can ever be expected when an unrepaired CP is repaired after adolescence. So there is a dynamic tension between the need for early palatal repair to facilitate good speech and the probable need for preservation of unoperated hard palate to allow normal maxillary growth. Schweckendiek performed soft palate closure at the time of lip repair and obturated the hard palatal cleft. Surgical closure of the latter was done after puberty (34). Independent evaluation of the results by Bardach et al. showed poor speech outcomes (40).

Given the availability of fairly predictable orthognathic surgery (Le Fort I advancement osteotomy) to treat deficient facial growth and thus yield an overall good skeletal outcome, and the apparent inability of speech therapy and secondary velopharyngeal surgery to eliminate the stigmata of CP speech following delayed palate repair, we believe in one-stage closure of hard and soft palate in time to produce a reasonably normal speech apparatus by the time that

infant's practicing of speech techniques begins in earnest, and thus aim to accomplish palate closure between 9 and 12 months of age.

ALVEOLAR BONE GRAFTING

The timing for definitive surgical management of the alveolar cleft remains unresolved. The essential goal of primary alveolar bone grafting is to inhibit substantial transverse maxillary collapse and occlusal distortion. However, secondary bone grafting of the maxilla in the mixed transitional dentition stage has become a well accepted procedure in the eventual dental rehabilitation of the cleft patient (41). Justification for closure of alveolar clefts includes stabilization of the alveolar arch, provide a template for orthodontic movement of the lateral incisor into the cleft, and enhance the hypoplastic pyriform region for definitive nasal correction. Recent advances with preoperative orthopedics and the employment of gingivoperiosteoplasty have resulted in some bone formation within the cleft margins.

Primary alveolar bone grafting performed at the time of CL closure is controversial due to the possible adverse effects on ultimate maxillary growth. However, secondary bone grafting appears to have the most widespread acceptance at the current time (42). Chronologically, this is performed at the stage of mixed dentition between 7 and 11 years of age to take full advantage of orthodontic therapy and resultant maxillary growth. Various donor sites have been proposed including the iliac crest, calvarium, rib, and anterior tibia (43). Our preference is to use cancellous bone, since it possesses osteogenic potential and will frequently survive even with mucosal dehiscence. It has been our experience that the optimal method of obtaining cortico-cancellous bone for reconstruction of the alveolar cleft is percutaneous harvest with a Craig biopsy needle, which allows this to be an outpatient procedure.

CLEFT PALATE REPAIR

A two-flap repair with an IVV and muscle transposition, much as described by Cutting, is frequently used. While the Furlow double Z-plasty has shown excellent results, true antero-posterior lengthening of the Z-plasty does not convincingly occur when there is lateral tension: in our hands, closure of the Furlow procedure often requires extensive lateral dissection, which risks compromising vascularity beyond that produced by the mucosal Z incisions alone.

By comparison, the two-flap repair with IVV preserves the mucosa and in our opinion allows more complete repositioning of all of the muscle. However, it does require more extensive muscle dissection, with more division of attachments of muscle to mucosa. It may be that the good functional results achieved with both the Furlow procedure and Cutting's IVV derive mostly from the excellent retro-positioning of the levator musculature to a fully transverse lie in the most posterior part of the soft palate, where it is arguably more mechanically efficient at producing velopharyngeal closure.

Procedure

After induction of general anesthesia, a throat pack is placed and intravenous cefazolin is given. The palate and vomer are infiltrated with xylocaine 0.5% with epinephrine 1:200,000 prior to sterile skin preparation and draping. Sterile prep and drape are probably of no value.

Complete Unilateral Cleft

The cleft margins are pared their full length. On the soft palate, we follow the visible junction line between oral and nasal mucosa, which, except near the uvula, is commonly seen on the oral surface 3 to 5 mm lateral to the apparent cleft margin. This will decrease closure tension on the more fragile nasal layer repair. Dissection of the oral and nasal mucoperiosteal layers follows. Tissue elevation is begun with a small curved-tip periosteal elevator. Occasionally, narrow clefts can be closed via the medial incision only. In this setting the heavy Blair elevator works well to follow the small elevator on the oral layer and can easily raise the flaps out to the alveolar ridge. However, we readily create lateral relaxing incisions and continue them along

the alveolar margin if more transverse laxity is needed, usually completely freeing the long flaps anteriorly to the alveolar cleft. If the palate is intact anterior to the incisive foramen, a Dorrance incision can be used anteriorly, along the gingiva, which avoids the confluence of incisions near the incisive foramen that accompanies the VWK repair. Complete hemostasis should be obtained along the flap margins at this time, which can be facilitated by elevating the anterior and lateral flap margins with the cautery after knife incision. Care should be taken to preserve the greater palatine neurovascular bundles.

The next task is to generate sufficient tissue laxity to allow easy movement of the flaps to the midline. This is done in three steps. First, the hook of the hamulus is exposed and the tendon of the tensor veli palatini completely divided. Next, the greater palatine neurovascular bundle is gently but firmly stretched out of its foramen using spreading, typically with scissors. Finally, an incision is made on the mucoperiosteum of the elevated oral flap immediately lateral to the vascular pedicle and the tissues delicately spread.

The nasal layer is then elevated out to the nasal side wall. After the first few millimeters, this is typically a blind dissection. On the medial side the exposed side of the vomer is elevated submucoperichondrially.

Assuming the vascular pedicles remain intact, one can now proceed with dissection of the levator musculature, separating it from the nasal and oral layers, usually in that order. Should one or both of the pedicles have been damaged, one should abandon the plan for IVV and instead proceed to approximate the undissected sides of the soft palate. This approach maintains nourishment to the hard palatal mucoperiosteal flap via oral mucosal vasculature and should prevent flap necrosis. If there is doubt as to the integrity of the vascular pedicles, look for the slight but distinct twitch of the oral mucoperiosteal flap with each pulse.

As the levator muscle lies against the nasal mucosa, the muscle should be dissected directly off the mucosa. One may leave some of the muscle against the oral mucosa when dissecting this surface, as this leaves behind tensor muscle and palatopharyngeus, which are antagonistic to levator function. Separation of the muscle from the posterior free margin of the hard palate should now be completed, if this has not previously been done. The muscle is then teased free from its connections to the side wall. Muscle dissection should be extended only as far posteriorly as necessary to allow the muscle bellies to be rotated medially 90° so that they can be overlapped in the posterior midline.

Adequate mobilization should be verified by pulling medially on the oral mucosal layer near the junction of hard and soft palate. If this does not move sufficiently, re-check the tensor tendon and then the mobilization of the vascular pedicles.

Repair of the palate is then done in three layers. The nasal layer is closed, generally from the uvula anteriorly. At the hard palate, the nasal layer is closed to the vomerine flap. The levator muscle flaps are then overlapped in the posterior midline (1 cm is routinely achievable) and secured together with interrupted sutures. The oral layer is closed; the hard palatal portion is closed with alternating simple and horizontal mattress sutures, the latter mattresses down to the nasal layer to close dead space. The lateral margins of the oral flaps are loosely tacked down to the cut alveolar margins, which usually leaves exposed bone laterally.

The repair has not addressed the alveolus. While this may be closed at the same procedure if there is good alignment of the greater and lesser segments, there is inconsistent bony union following this procedure; whereas separate closure later, in mixed dentition, allows more predictable bone graft survival in an unscarred bed.

Postoperative Care

The throat pack and mouth gag are removed, and a two to zero silk suture is passed through the tongue tip, tied into a loop, and taped to the cheek under no tension. This allows passive protrusion of the tongue to open a swollen and narrowed airway should it be required. The suture is removed the next morning. Elbow splints are placed and will be left for two weeks. Continuous blow-by mist with room air is used overnight to prevent hardening of the expected palatal drainage. A clear liquid diet is begun on post-operative day 1, and the child is discharged when adequate oral intake is demonstrated.

COMPLETE BILATERAL CLEFT PALATE REPAIR

Dissection is the same as for the unilateral cleft, except for the obvious fact that there are two palatal shelves that require nasal layer dissection and that the vomerine flaps are elevated bilaterally. Also, the residual oro-nasal fistula is smaller if the anterior tips of the oral and nasal mucoperiosteal flaps are closed to the corresponding flaps of the premaxilla. To achieve this, continue the incision of the vomerine margin anteriorly into a Y split along the premaxillary edges. Continue elevation of the vomerine flaps anteriorly onto the premaxilla so they can be sutured across the clefts to the nasal layer flaps. Sew the medially repositioned oral mucoperiosteal flaps to the premaxillary mucosa. Post-operative care is as for unilateral CP.

VELOPHARYNGEAL INSUFFICIENCY

The frequency of the eventual development of completely normal speech following CP repair remains variable, with reports ranging from less than 50% to 95.8% (44). Currently, the plastic surgeon has at his/her disposal various tools to complement their clinical observations of nasality. The nasendoscope appears, in our opinion, to possess the most significant capacity for evaluation of velopharyngeal anatomy and function. Based on the results of this examination, the surgeon can make an educated plan for surgical correction via either a superiorly-based pharyngeal flap or a sphincter pharyngoplasty.

EPILOGUE

While good outcomes of appearance and function following surgical treatment of CL and CP have sometimes been achieved by surgeons for nearly a century, the average level of accomplishment by all cleft surgeons is still improving, as techniques continue to evolve and disseminate and as studies continue to point the way to the best treatment protocols. The long period required to evaluate a given protocol suggests that we may not know the best surgical approach for perhaps several decades. However, the rapid information-sharing now available will allow better prospective studies. Already, the overall high level of surgical training available and the widespread use of team care mean that children with clefts in advanced countries and in a growing number of developing countries are able to live normal productive lives.

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18. This is not strictly true. On a convex surface an equilateral triangle will have all 3 angles greater than 60°.
19. Perhaps this works because the non-cleft side often has a slight concavity (negatively-curved strip) supradjacent to the white roll, causing the caliper measurement (which measures Euclidean distances) to underestimate the amount of lip height needed along the skin surface when this number is transferred to the (sometimes planar or convex) cleft side. Another surgeon may or may not find this extra millimeter necessary.
20. Children are very adaptable and will quickly adjust to splinting. It may be the parents who need to be managed. Leaving on the splints avoids “game-playing” and subsequent frustration of parent and child. When necessary, a shoelace tied across the back between grommets on the proximal ends of the splints will foil even the most ingenious Houdini.
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11

Management of Secondary Maxillary and Nasal Deformities in Adolescent Cleft Lip and Palate Patients

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INTRODUCTION

Management of secondary maxillary and nasal deformities in the adolescent cleft patient can present a formidable surgical challenge. The functional as well as aesthetic needs of each patient must be given careful consideration. A team approach can optimize the preoperative planning, procedure selection, and postoperative management. Team members include Speech and Language Pathologist, Orthodontist, Dentist, Prosthodontist, and Psychologist. The author's management protocol for surgical correction of deformities of the midface and nose in adolescent patients is presented. This has been successfully used at Children's Healthcare of Atlanta for over 17 years.

MANAGEMENT OF CLEFT/MAXILLARY DEFORMITIES

Approximately 15% to 20% of adolescents with unilateral or bilateral cleft lip and/or palate will have some degree of maxillary hypoplasia. The underdevelopment of the maxilla may be noted soon after birth and is often progressive. In many cases correction requires a combined orthodontic and surgical approach. The results of maxillary hypoplasia include Class III malocclusion with abnormal wear and tear on the teeth, masticatory difficulties, nasal and nasopharyngeal airway obstruction, and aesthetic facial deformity. Dental deformities often accompany the maxillary deformities and can include missing dental units, supernumerary teeth, hypoplasia of dental enamel, and deformed teeth (Fig. 1).

I consider the nose and maxilla to be a single aesthetic unit when considering planning for surgical correction. The position and size of the lower jaw and chin are also assessed aesthetically. The midface appears to be concave rather than convex, which decreases nasal base projection, as well as tending to make the ala somewhat splayed. The upper lip is poorly supported and there is often inadequate dental show. The underlying skeletal deformity magnifies the cleft nasal deformity (Fig. 1). As part of the treatment planning process cephalometric radiographs, facial and dental photographs are taken. The cephalometric data can be digitized to allow for precise treatment planning (1,2). Computerized facial imaging which simulates both the maxillary procedure and the subsequent nasal surgery is often helpful in explaining the treatment challenges and goals to the patient and family (3,4). The ultimate aesthetic and functional goals are discussed with the patient and their family. Each step in the process, including dental and orthodontic preparation, surgical correction of the maxillary and nasal deformity, finishing orthodontics, prosthetic dentistry, and if necessary psychological evaluation are presented. The patient, family and multidisciplinary team have to agree on the process, timeline, and goals prior to the start of treatment. Reasonable expectations are discussed and agreed on by all participants. I prefer to correct the maxillary deformity first, thus establishing a bony base for future definitive nasal reconstruction. At least nine months are allowed between maxillary advancement and the definitive internal and external nasal

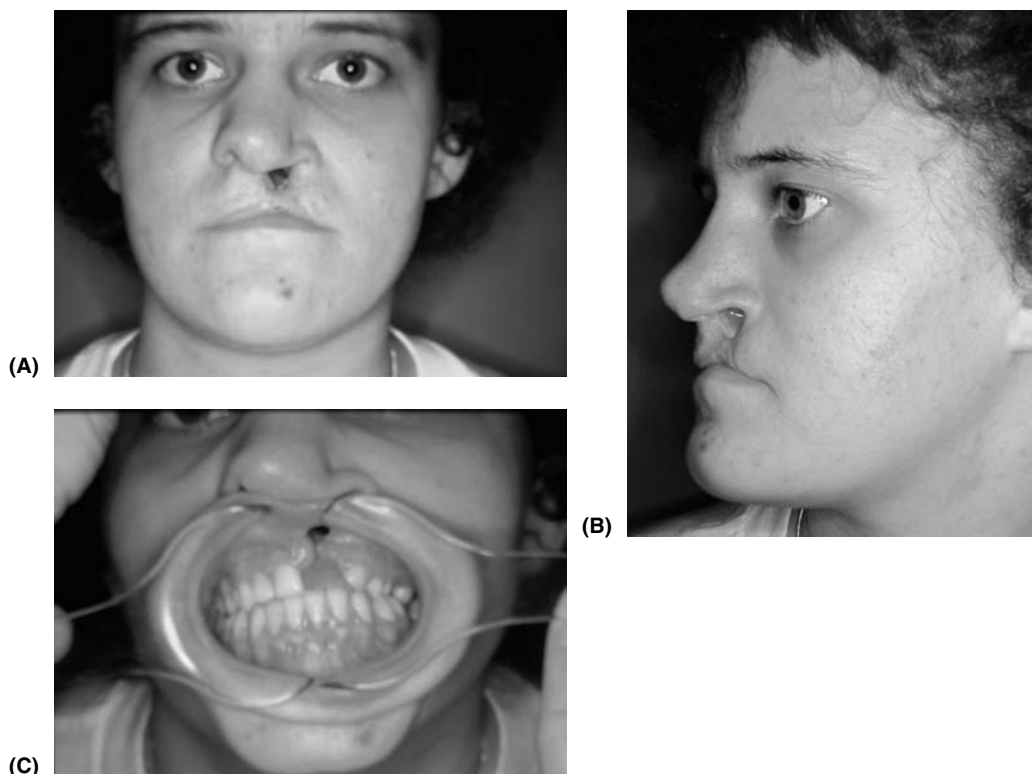


FIGURE 1 (A–C) Fifteen-year-old patient when she presented after repair of unilateral cleft lip and palate elsewhere. Note oral nasal, oral cutaneous fistulas, alveolar cleft, maxillary hypoplasia, and severe cleft nasal deformity. Note absence of central and lateral cleft side incisors.

reconstruction. I do not recommend simultaneous nasal reconstruction at the time of maxillary advancement.

Maxillary Advancement

Planning for surgical correction of severe maxillary hypoplasia in the cleft patient has to begin early in life. Ideally, I perform a gingivoperiosteoplasty at the time of lip repair. In some cases this will suffice and avoid alveolar bone grafting in the future. Even when there is not enough alveolar bone after gingivoperiosteoplasty, it often helps to align the maxillary segments and avoid chronic oral/nasal fistulas. If a child does need alveolar bone grafting, this is coordinated with his/her orthodontist. Orthodontic preparation for bone grafting includes maxillary expansion and, if necessary, aligning the teeth adjacent to the cleft. My bone graft technique includes a minimal incision, utilization of a bone harvesting mill, and packing the donor site with a bupivacaine impregnated resorbable implant (5). I have found that this routine makes the procedure relatively painless and allows for 23 hour stay in the hospital and resumption of normal activity within two to three days. Orthodontic treatment is resumed within three to four weeks after the alveolar bone grafting, but the palatal expansion device is left in place for four to six months to allow bony consolidation. Maxillary advancement can proceed after 12 to 18 months of bone consolidation. This approach allows for a single piece maxillary advancement rather than multiple segments with simultaneous bone grafting to the alveolar cleft at the time of maxillary advancement.

Orthodontic preparation for maxillary advancement begins approximately one year prior to the time of the scheduled surgery (6). This includes aligning and leveling the occlusion, as well as any dental extractions that may be necessary to get proper dental spacing.

Approximately two weeks prior to the planned maxillary surgery heavy orthodontic wires are placed, along with hooks. At the same time, mounted models are taken for model surgery (7,8). During the same visit, a complete speech and language evaluation, including nasal endoscopy, air flow, and articulatory studies, are performed. This allows us to identify the patients at high risk for velopharyngeal incompetence after maxillary advancement. The patient and parents are informed, if this is the case, preparing them for the possibility of a pharyngoplasty or pharyngeal flap in the future if velopharyngeal incompetence does occur. Model surgery is performed using articulated models, to achieve optimal occlusion, and intraoperative splints are made.

Special considerations at the time of surgery include preoperative antibiotics and intravenous steroids. Hypotensive anesthesia is preferred and all patients are asked to donate a unit of autologous blood or donor-directed blood. The preoperative nasal endoscopy will reveal the location and portal size if a previous pharyngeal flap is present. The design of the osteotomies can vary according to the patient's aesthetic needs (9). In patients with hypoplastic malar regions, the osteotomies are made quite high, just under or including a portion of the inferior zygomatic arch under the inferior orbital nerve, in order to get maximal bony fullness with the advancement. Vertical measurements are taken from the medial canthus to the incisor brackets on both sides to assure that the occlusion stays level and to determine the vertical dimensions. Care is taken to dissect out the mucosa of the medial and inferior nasal cavities and to protect the nasotracheal tube. Once the osteotomies have been completed, the maxilla is mobilized with a combination of Rowe disimpaction forceps and Wolfe disimpaction devices. If there is a restrictive pharyngeal flap, it is taken down at this time to allow full mobilization of the maxilla. In many cases scar tissue will make mobilization difficult requiring considerable force. The color of the gingival should be carefully monitored during mobilization to avoid devascularization. The patient is then put into the pre-fabricated occlusal splint, from the model surgery, and then into intermaxillary fixation. Once the patient has been put into intermaxillary fixation with both rubber bands and wires, the final vertical dimensions are determined. This is done from preoperative analysis and photographs. Bony interferences are taken down if an impaction is required. If necessary, the Golden Rule, as popularized by

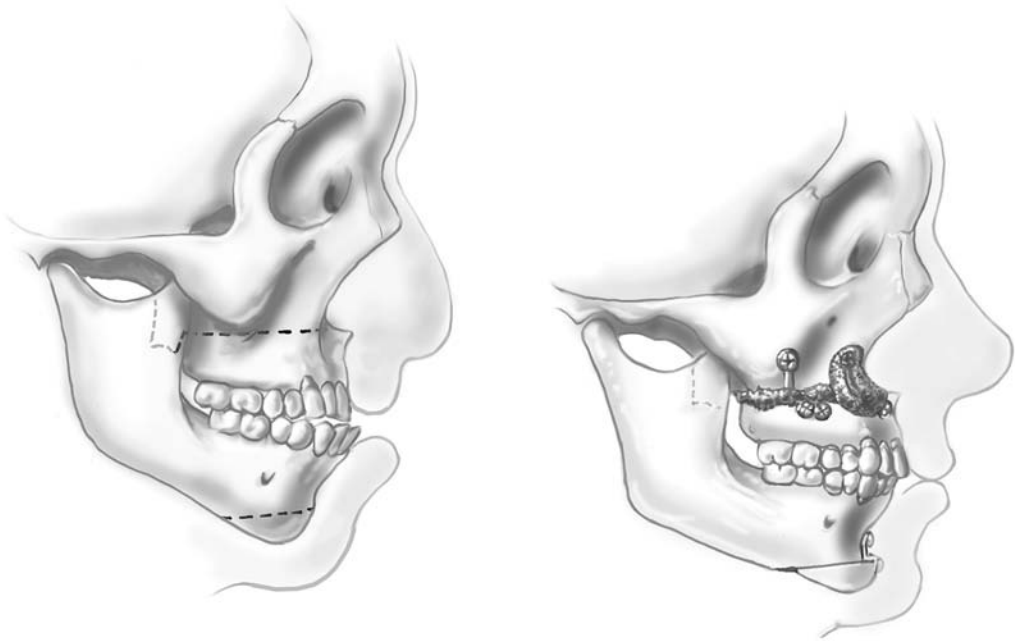


FIGURE 2 Drawing of maxillary advancement and horizontal osteotomy of mandible. Note placement of rigid fixation plates at medial and lateral buttress and bone grafting of advancement gap.

Rickett's, serves as a very useful guide to determine the optimal aesthetic proportions. In most cases, it will be necessary to bone graft the gap between the proximal and distal segments (Fig. 2). I prefer to use iliac crest bone graft for this. I have found that the easiest and least painful donor site simply involves going back through the old bone graft incision, if one is present, extending it slightly, retracting the soft tissues medially, and then harvesting the inner table of the iliac crest with a reciprocating saw and osteotomes. A very large cortical cancellous graft can, thus, be obtained. Because the iliac crest height has not been violated, this does not leave a hip contour defect. The wound is closed in the same fashion as done for alveolar bone grafting. If a substantial graft has been obtained, a drain is left overnight to prevent hematoma formation. Next, the gap between the proximal and distal segments is measured. I prefer to use specialized, extreme plates manufactured by the KLS Company. These plates are pre-bent to approximate different advancements from 7 to 15 mm and are 2 mm thick, making them extremely stiff and resistant to postoperative bending (Fig. 3). Screws of 2 mm diameter are used to get maximal bone grip. Proper placement of these plates is critical to avoid a poor occlusal result. Typically, the medial and lateral buttresses represent the strongest bone available for fixation and it is here that the best application of the rigid fixation can be made (Figs. 2 and 3). The plates need to be precisely bent to the contours of the maxilla to ensure occlusal stability, when using the pre-contoured plates this usually requires only minor adjustments. Two plates per side are utilized. At this point, if there is any doubt regarding the integrity of the previous alveolar bone graft, it is wise to expose the area and re-graft it from above. If there are very large turbinates, they can be trimmed at this point. If an impaction is contemplated in the vertical dimension, it may be necessary to remove a segment of septum to

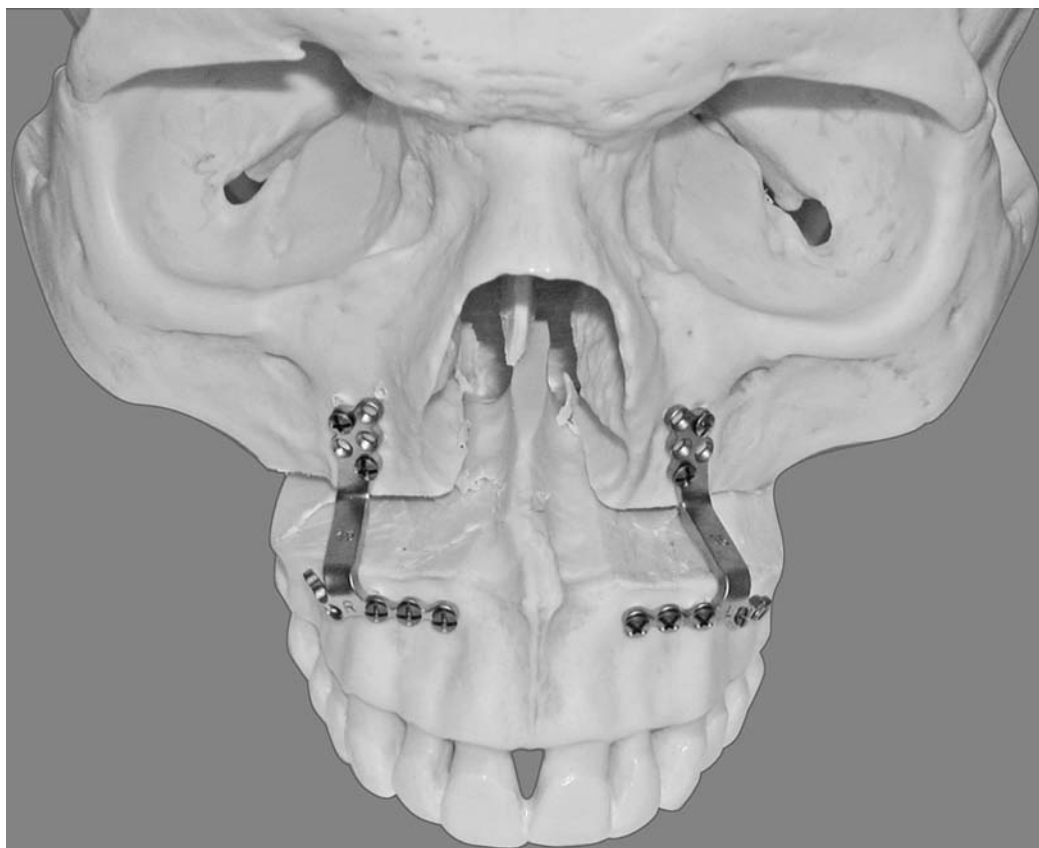


FIGURE 3 Skull model with medial buttress fixation with “extreme,” 2 mm thick, pre-contoured plating system.

prevent it from warping during the impaction, causing postoperative airway obstruction. Once these maneuvers have been completed, the patient is taken completely out of maxillary fixation and, with the mandible in centric relation, with gentle posterior pressure, the occlusion is checked. The occlusion should fit passively into the occlusal splint. If it does not, it indicates that either the plates were not bent precisely, causing torque on the maxilla, or the condyles were not seated at the time the patient was put into maxillary fixation. In either case, reapplying the intermaxillary fixation is required, with removal of all hardware. All bony interferences should be resected, and the plates contoured to fit precisely against the bone. If the occlusion is acceptable at the end of the rigid fixation phase of the procedure, the patient is put into simple 8 oz elastics with slight anterior maxillary pull, the occlusal splint is wired to the orthodontic wire. If the patient has had an impaction, a cinching suture is placed along the base of the nose to prevent widening of the ala. These wounds are irrigated with antibiotic solution and closed with resorbable sutures.

Postoperatively, the patient is extubated in the operating room and put into an upright position with ice packs. Intravenous steroids are continued for two days after surgery. The patient is put immediately on a clear liquid diet and then advanced to a soft diet as tolerated. The hip drain is removed within one to two days, depending on the amount of drainage, and the patient is discharged. At the first week's postoperative appointment, the patient is shown how to change the rubber bands and is allowed to remove them to eat. The occlusal splint is removed after two weeks. A soft diet is instituted for six weeks. A follow-up orthodontic appointment is made within four weeks of the surgery.

Potential problems with the surgery include relapse of the maxilla to a variable degree. This can be prevented by continuing to have slight anterior pull on the light elastics or consideration can be given to a reverse pull face gear to exert tension on the maxilla. Care should be taken not to exert too much pull on the maxilla during the early healing period, since it is possible, through continuous movement and elastic tension, to actually begin to dislodge the fixation. After maxillary stability has been achieved, finishing orthodontics is instituted. If prosthetic rehabilitation for missing teeth is necessary, I recommend waiting at least 9 to 12 months after the surgery to allow full healing of the surgical site. After that time, the orthodontist, prosthodontist, and implant surgeon can decide the best strategy for final rehabilitation of the dentition (Figs. 1,4, and 5). If postoperative velopharyngeal incompetence is detected the patient is monitored by the Speech and Language Pathologist for six months. If no improvement is forthcoming a pharyngoplasty is performed.

SURGERY FOR SECONDARY NASAL DEFORMITIES

Final nasal reconstruction is typically done when skeletal maturity has been achieved. I prefer to wait to do the definitive rhinoplasty until after the maxilla is in its proper alignment. If a maxillary advancement is performed I wait 9 to 12 months before rhinoplasty. My approach to cleft nasal reconstruction includes consideration of both functional and the aesthetic needs of the patient. Computerized imaging is often utilized to help the patient visualize the idealized final appearance and to explore the patient's expectations preoperatively (3,4). Most patients with cleft lip and palate will have some degree of nasal airway obstruction (10–12). This is typically caused by nasal septal deviation, including the vomer, perpendicular plate of ethmoid, and the cartilaginous septum. Often, there will be compensatory turbinate hypertrophy contributing to the overall nasal airway obstruction. In addition, the nasal inlet may be obstructed by inspiratory collapse either at the lower lateral cartilage level or alar valve level. All these sites need to be taken into consideration. This surgery is carried out under general anesthesia and an open rhinoplasty approach is used in all cases (13).

Unilateral Cleft Nasal Deformity

The unilateral deformity has several characteristic components that must be considered during preoperative planning and surgical repair. These include nasal tip deviation toward normal side, posterior displacement of cleft side dome, alar buckling on cleft side, medial



FIGURE 4 (A–C) Patient in Figure 1 at 19 years of age, i.e., two years after completion of reconstructive procedures. She underwent alveolar bone grafting, lip revision, and closure of fistulas at 15. This was followed one year later by maxillary advancement and horizontal osteotomy of the mandible at 16. The final nasal reconstruction was at 17. **(C)** Demonstrates the final occlusion and prosthetic restoration of the absent central and lateral incisors.

crus retro displacement on the cleft side, septum and collumelar deviation toward the cleft side, and buckling of the lateral crura on the cleft side (Fig. 6). The unilateral cleft reconstruction involves a stair step collumelar incision and a rim incision, with exposure of the entire nasal pyramid (10,11). It is very important to be able to visualize the anatomy fully. The nasal septum is resected from between the two medial crura of the lower lateral footplates with mucosal flaps elevated on either side. At least 1.5 cm of dorsal septum should be kept intact. Often, the caudal septum is grossly deviated and must be resected. The deviated portions of the septum are completely resected, to be used as autologous



FIGURE 5 (A–F) Patient with bilateral cleft lip and palate, maxillary hypoplasia, nasal deformity, and lip imbalance. (A–C) Demonstrate appearance and occlusion at 13 years of age. (D–F) Demonstrate final appearance and occlusion. Patient underwent maxillary advancement and horizontal osteotomy of mandible at 15. At 17 she underwent nasal reconstruction and dermal fat grafting to the lip and sub nasal areas. She also had prosthetic dental rehabilitation.

cartilage grafts. If the perpendicular plate of ethmoid and vomer are involved, they should also be resected at that time. Once this has been accomplished, the mucoperichondrial flaps are sutured to each other using resorbable sutures. A conservative inferior turbinectomy is advocated to prevent nasal crusting or drying. Once the nasal airway has been established, the height of the dorsum is set and, if necessary, rasping is done to take down any dorsal irregularities or humps. Care is taken to create junction tunnels between the upper lateral cartilages and the septum. Next, the bony vault is addressed. If the dorsum has been taken down or if the dorsum is wide, lateral osteotomies are carried out with a 2 mm osteotome to close the open roof deformity and to decrease the nasal width. Next, the mid-vault is addressed. If there is significant dorsal septal deviation, this can be camouflaged with spreader grafts (13). These are placed on the concave side and should extend across the nasal valve (Fig. 6). They are secured with resorbable suture. Next, the nasal tip is addressed. The nasal tip deformities can be quite severe and include asymmetry, hypoplasia of the cartilages, and displacement of the cartilages. The cephalic margin of the both lower lateral cartilages are usually trimmed in order to provide a more refined nasal tip and then over-grafting is carried out, on the cleft side, to make up for any cartilage deficiencies (Fig. 6). A collumellar strut graft should be utilized if the caudal septum has been removed (Fig. 6). The strut graft is sutured to the medial crura of the lower lateral footplates with resorbable sutures. Finally, a variation of several types of tip grafts can be used to give tip definition. These include shield grafts and stacked Peck-type grafts (14). Packing is not used and the external incision is closed with synthetic sutures. If the alae are wide or

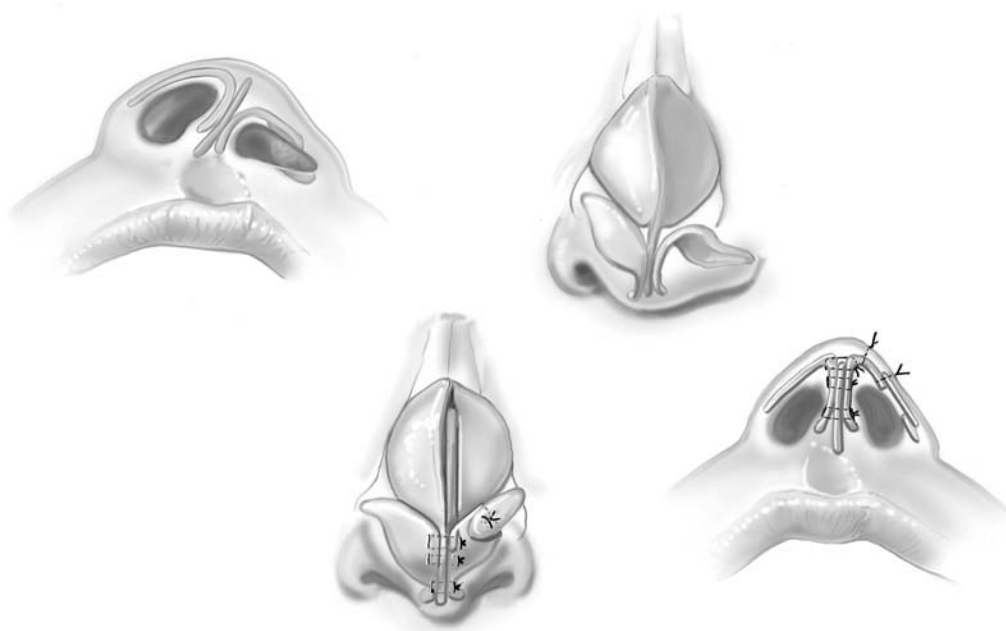


FIGURE 6 Drawing of unilateral cleft nasal deformity and repair. Top row demonstrates typical deformities from submental vertex and head-on view. Bottom row depicts correction with autologous septal grafts, including alar spreader, columellar, and alar onlay grafts.

asymmetric, they are addressed at this point. These should be addressed with either alar reduction, going along the floor of the nose, or, more commonly, by repositioning the cleft side ala. Typically, the ala tends to be displaced laterally. In order to reconstruct the floor of nose, a Y to V advancement of the lateral ala is done. This can be done with an 11 scalpel and matched to the contralateral side without much difficulty. Adhesive strips are applied, along with a thermoplastic dressing. The patient is discharged on oral antibiotics and pain medication, they are allowed to shower. We have found that a short course of postoperative steroids can also be quite helpful in reducing swelling. All permanent sutures are removed at one week as is the splint, saline drops are started to help clear out the airway.

Bilateral Cleft Nasal Deformity

The bilateral deformity is quite different from the unilateral cleft deformity (Fig. 7). The bilateral deformity includes a foreshortened nasal tip, a short columella, bifid but symmetric tip cartilages, a wide nasal dorsum, and low alar domes, buckling of the lateral crura, and variable degrees of septal displacement (10,12). If the columella is adequate, the incisions can be made with a stair step design. In most adolescent patients there has already been surgery to lengthen the perceived lack of columellar length. If the columella is short, the bilateral lip reconstruction scars can be harvested as small forked-flaps to lengthen the columella. Alternatively, wide undermining of the skin envelope will often preclude the need for columellar flaps if adequate structural support is provided. The rest of the exposure is standard external rhinoplasty exposure (13). If there is a component of airway obstruction, this can be addressed in the same fashion as in the unilateral deformity. Often, there is fibro fatty tissue between the splayed medial crura, which should be removed to give maximal tip refinement. The lateral crura are then mobilized from the soft tissues with sharp scissor dissection. Next, the nasal dorsum is set. This can include taking down any type of hump or doing lateral osteotomies to decrease the overall nasal width. If the dorsum is adequate but too wide, lateral osteotomies via a pyriform aperture approach using a 2 mm osteotome are

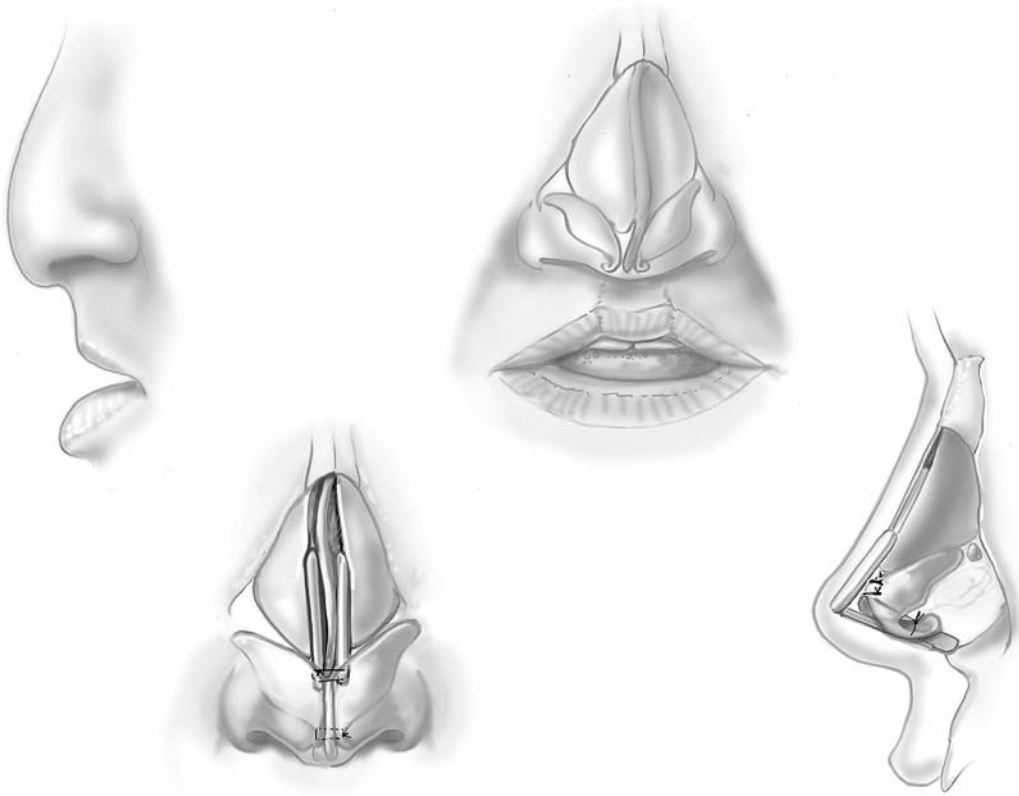


FIGURE 7 Drawing of bilateral cleft nasal deformity and repair. Top row demonstrates blunted lateral appearance with short columella and anterior posterior view of cartilaginous deformity. Bottom row depicts lengthening of nose with combination of extended spreader grafts, columellar strut, and shield graft to tip.

carried out. If necessary, the nasal dorsum can be augmented with either on lay septal grafts or, more commonly, with extended alar spreader grafts that are placed above the level of the dorsal septum to give dorsal definition (Fig. 7) (13). These are sutured onto the dorsal septum and to each other with resorbable sutures. The upper lateral cartilages are then suspended to each other. The nasal tip then needs to be lengthened. This can be done through a combination of techniques, depending on the severity of the deformity. The medial crura are sutured to each other, as are the footplates, essentially walking length up the nasal columella. If this is not adequate or if the nose is extremely short, tip projection and length must be added. If adequate septal cartilage is available, this is ideal. This can be done by a series of shield grafts placed with the wide end downward and also along the nasal columella to essentially extend the tip downward and forward, (14). It is important that these grafts all be secured to each other and to the underlying dorsum with resorbable suture. A columella strut graft can be quite helpful to secure this construct (Fig. 7). Wide undermining of the nasal skin is advocated in order to have adequate soft tissue length to cover the grafts. Occasionally, septal cartilage is not present or is inadequate. A conchal graft can serve as an excellent substitute. The perichondrium is left on one side of the graft and the graft is trimmed to resemble an L-strut. It is then sutured to the underlying medial crura and nasal tip, leaving graft projecting to extend the tip. Conchal grafts can also be stacked on this construct in order to increase tip projection and length. All intranasal incisions are closed with resorbable sutures and the external incisions with non-dissolvable sutures. The ala will often need to be adjusted and a Y to V alar advancement medially

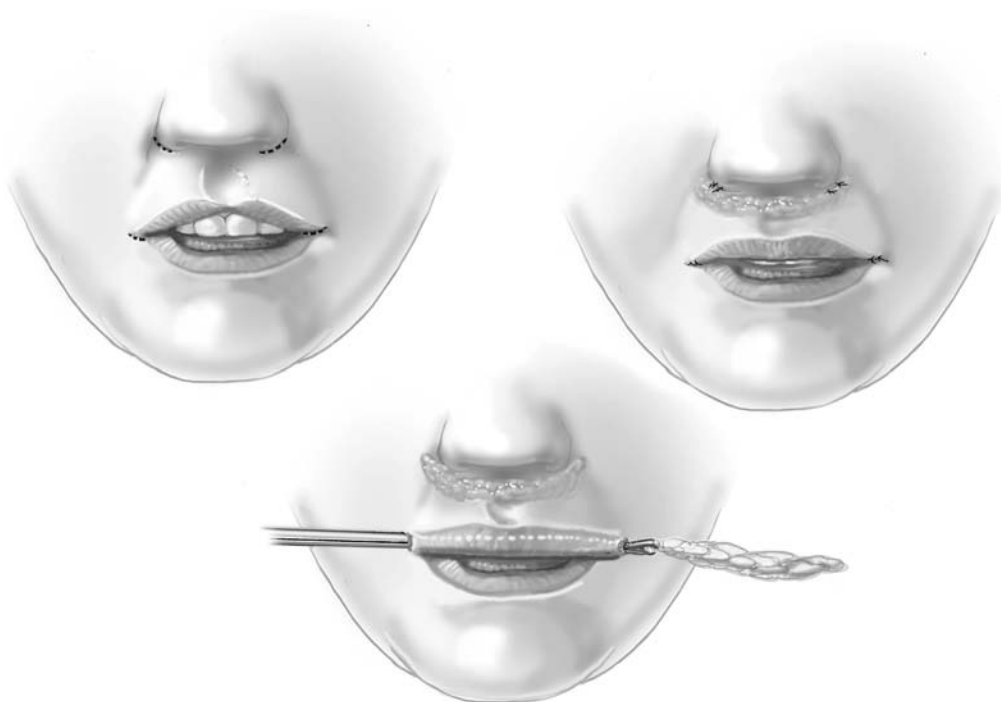


FIGURE 8 Drawing depicting incisions and placement of dermal fat grafts for lip and paranasal augmentation.

is an excellent maneuver to decrease alar width. Postoperative care is the same as with an unilateral deformity.

Adjuvant Techniques

Adjuvant techniques are sometimes helpful to achieve the best final aesthetic result. Occasionally, the cleft patient with maxillary hypoplasia will have an imbalance in chin position or size. Genioplasty can be useful in achieving an optimal aesthetic result and is done at the time of maxillary advancement (Figs. 2 and 5) (15). Lip revision is often necessary and can be done at the time of rhinoplasty. With some patients there is an imbalance between the upper and lower lip or paranasal flatness despite a Class I occlusal relationship after maxillary advancement. In those cases, a dermal fat graft can be a valuable adjuvant measure (Figs. 5 and 7). Typically, the dermal fat graft is harvested from the suprapubic area. If the lip is to be augmented, a small incision is made on either side of the defect along the free border of the lip and deep subcutaneous dissection is carried out with sharp scissors. An adequate tunnel must be created and should be in the shape of the graft. The graft is trimmed to fit, leaving plenty of dermis to adhere to and pulled through the subcutaneous tunnel (Fig. 8). If the paranasal areas or sub nasal areas are still inadequate despite normal bony projection, the dermal fat graft is tunneled through the Y to V incisions for augmentation. Approximately 30% resorption can be expected, so these grafts should be made somewhat larger than anticipated in order to allow for shrinkage.

CONCLUSION

Surgery of the midface and nose are valuable tools in completing reconstruction for adolescents and adults with bilateral cleft lip and palate. Often, these are the last surgeries needed for their reconstruction. Careful preoperative planning, computerized imaging, and a team approach to both the orthognathic and aesthetic surgery can yield gratifying results.

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12 Craniofacial Clefts

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INTRODUCTION

Rare craniofacial clefts are one of the most disfiguring facial anomalies. They have unusual presentations and seem to defy description; however, most craniofacial clefts occur along predictable embryologic lines (1–4).

EPIDEMIOLOGY

The exact incidence of rare craniofacial clefts is unknown; however, they have been estimated at 1.4 to 4.9 per 100,000 live births. Comparing rare craniofacial clefts to common clefts, their incidence seems to approximate 9.5 to 34 per 1000.

ETIOLOGY

The majority of rare craniofacial clefts occur sporadically and their etiology is unknown. There are, however, familial patterns of inheritance for rare craniofacial clefts in Treacher–Collins and Goldenhar syndromes. Amniotic band syndrome and environmental factors have also been implicated in the process of facial clefting.

THEORIES OF FACIAL CLEFTING

Two theories have been developed to describe facial cleft formation. The first theory states facial clefting is caused by a failure in the facial fusion processes. The second theory suggests that there is a failure of mesodermal migration and penetrance of neuroectoderm. Neither theory has been proved and the precise nature of the proposed mechanism in the formation of rare craniofacial clefts remains a mystery.

PATHOGENESIS

The stapedial artery is present during fetal development and gives the blood supply for the first and second branchial arches. Experimentally induced localized hemorrhage of the stapedial artery in laboratory animals has induced facial clefting. Depending on the magnitude and timing of the local tissue destruction, varying degrees of malformation are observed.

CLASSIFICATIONS

Over the years several classifications have been created to describe the rare craniofacial clefts. They include: American Association of Cleft Palate Rehabilitation Classification by Harkins et al. in 1962, Karfik Classification in 1966, Van der Meulen and Associates Classification in 1983, Median Tissue Deficiency Classification by DeMyer in 1964, and Median Excess Tissue Classification by DeMyer in 1967. In 1973 Tessier presented a classification of craniofacial clefts that is the most complete and has withstood the test of time. The unique classification is based on

the extensive personal experience of Dr. Paul Tessier, which has enabled the terminology to remain uniform.

TESSIER CLASSIFICATION OF CLEFTS

General

Tessier's classification of craniofacial clefts (Figs. 1 and 2) has been the most enduring and clinically relevant classification, linking clinical observations to underlying skeletal deformity. The clefts are numbered from 0 to 14 and follow well-defined "time zones." The eyelids and orbits defines axis and divides face into upper and lower hemispheres. The facial clefts are numbered from 0 to 14. Clinically observed combinations include: 0 and 14, 1 and 13, 2 and 12, 3 and 11, 4 and 10, and 5 and 9. Tessier cleft numbers 6, 7, and 8 are the lateral most craniofacial cleft.

Number 0 Cleft

1. *Soft tissue characteristics (widening or duplication).* This is a true midline cleft. There is a central incisor diastema, bifid labial frenulum, broad philtral columns, bifid nose, and duplicated nasal septum (Figs. 3 and 4).
2. *Bony characteristics (widening or duplication).* The maxilla is keel shaped with the anterior teeth angled medially, producing an anterior open bite and vertical maxillary hypoplasia. The cleft passes through the primary and secondary palate. There is a duplicated anterior nasal spine and septum, broad and flattened nasal bones, enlarged ethmoid and sphenoid sinuses, and orbital hypertelorism.
3. *Soft tissue characteristics (agenesis or hypoplasia).* This is a false facial cleft. It is associated with an absent philtral column, hypoplastic columella, primary and secondary cleft palate, nasal hypoplasia, vestigial nasal septum, and depressed nasal tip.
4. *Bony characteristics (agenesis or hypoplasia).* There is an absent nasal bone and septum, hypoplastic ethmoids, orbital hypotelorism, and eye deformities.

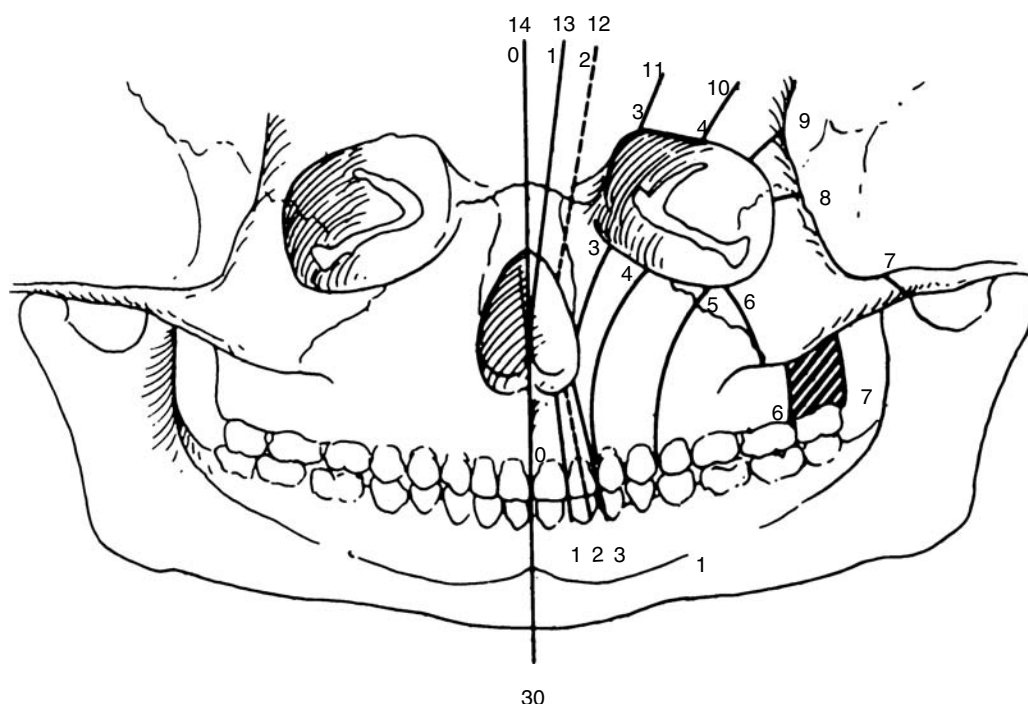


FIGURE 1 Tessier craniofacial clefts classification (bony).

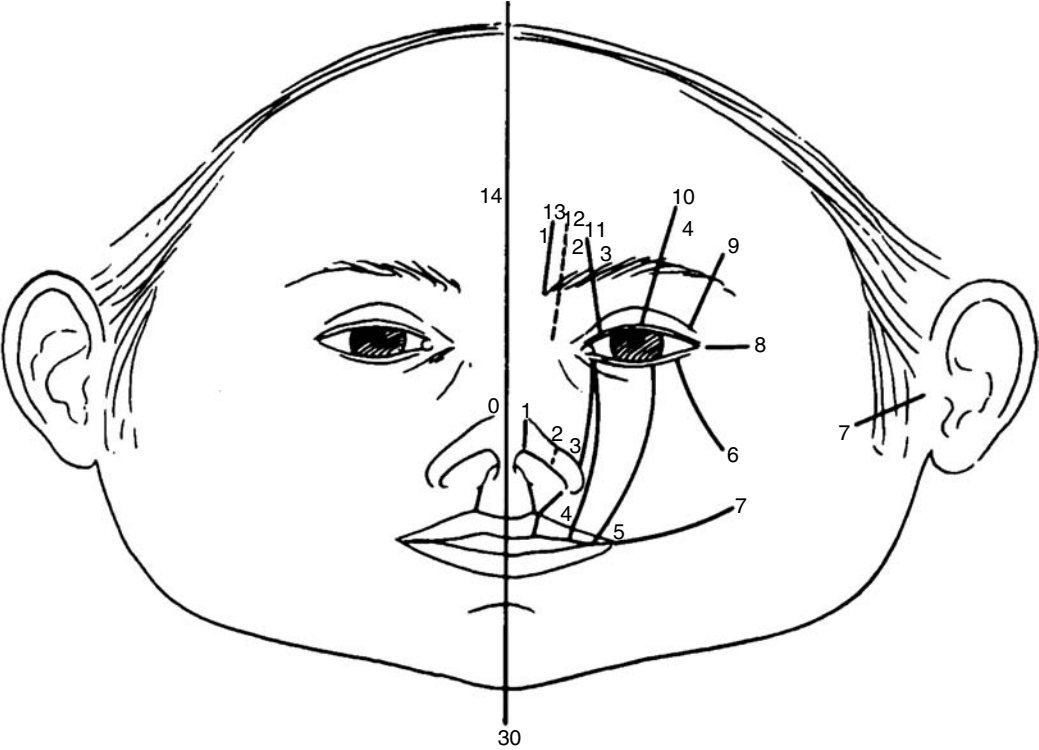


FIGURE 2 Tessier craniofacial clefts classification (soft tissue).



FIGURE 3 Tessier numbers 0/14 craniofacial cleft.

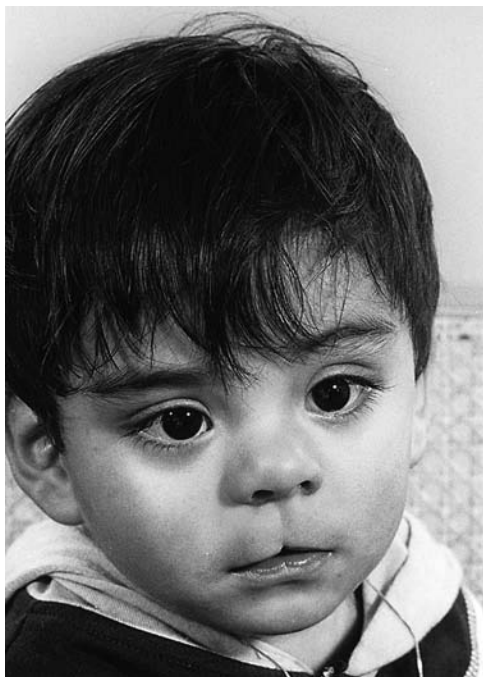


FIGURE 4 Tessier number 0 cleft with minimal hypotelorism. *Source:* Courtesy of Dr. H. K. Kawamoto.

5. *Soft tissue and bony characteristics (lower face).* The number 30 cleft is the caudal extension of number 0 craniofacial cleft. There is a median cleft between the central incisors and mandibular symphysis, neck deformities, absent hyoid bone, abnormal thyroid cartilage, atrophic neck muscles, bifid tongue, and ankyloglossia.

Number 1 Cleft

1. *Soft tissue characteristics.* The facial cleft is the same as the common cleft. In addition, the number one cleft is associated with a notched alar dome, broad columella, nasal tip, and septum deviated away from cleft and furrows along nasal dorsum. The cleft passes medial to the medial canthus, creating a vertical orbital distopia, and telecanthus (Figs. 5 and 6A,B).
2. *Bony characteristics.* The maxilla is again keel shaped with medially facing teeth and an anterior open bite. The cleft travels between central and lateral incisors, into pyriform aperture and through primary and secondary palate. The cleft then passes between nasal bone and maxilla, creating flattened nasal bones, ethmoid hypertrophy, and orbital hypertelorism.

Number 2 Cleft

1. *Soft tissue characteristics.* The cleft lip lies in the same region as the common cleft. There is alar cartilage hypoplasia, flattened lateral nose, and broad nasal dorsum. The cleft passes medial to the palpebral fissure, causing a laterally displaced medial canthus. Typically, the lacrimal drainage is normal (Figs. 7 and 8).
2. *Bony characteristics.* The cleft passes between lateral incisor and canine, through the primary and secondary palate and into pyriform sinus. The nasal septum is intact and deviated away from the cleft. The cleft then passes between nasal bone and maxilla, causing orbital hypertelorism. The maxillary sinus is typically intact, while the ethmoid and frontal sinuses are hypoplastic.

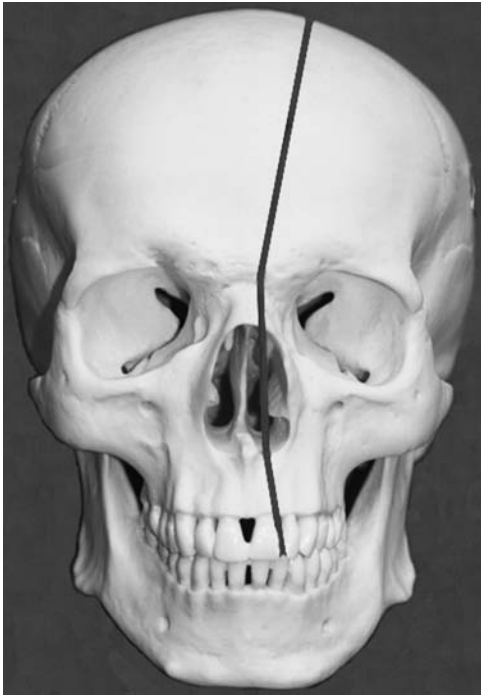


FIGURE 5 Tessier numbers 1/13 craniofacial cleft.

Number 3 Cleft

1. *Soft tissue characteristics.* This is the most common Tessier cleft. The cleft travels along the philtral column into the nasal floor. There is vertical shortening between ala and lower lid, creating a short nose and an upward pull of alar base. The lacrimal system and lower canaliculus are disrupted. Lower lid colobomas are common (Figs. 9 and 10A–C).



FIGURE 6 Tessier number 1 cleft with notch in medial alar rim. (A) Anterior view of cleft. (B) Inferior view of cleft. Source: Courtesy of Dr. H. K. Kawamoto.

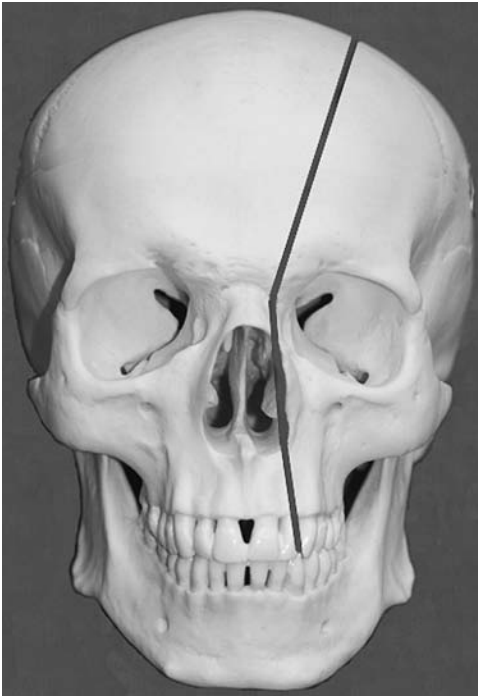


FIGURE 7 Tessier numbers 2/12 craniofacial cleft.



FIGURE 8 Tessier number 2 cleft with abnormality in lateral alar rim. *Source:* Courtesy of Dr. H. K. Kawamoto.

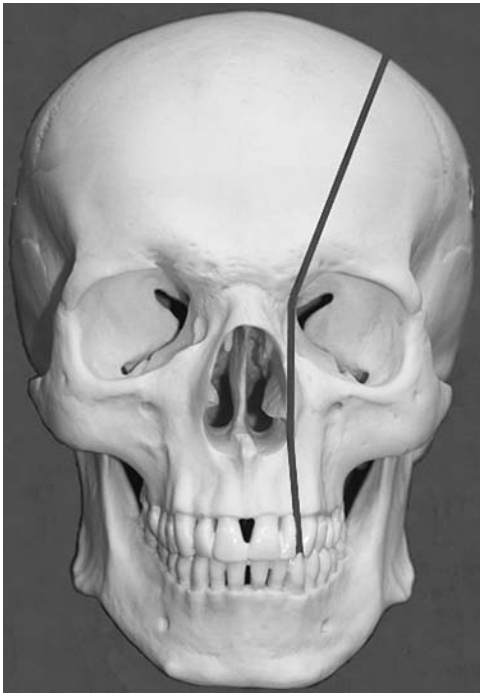


FIGURE 9 Tessier numbers 3/11 craniofacial cleft.

2. *Bony characteristics.* The cleft passes between lateral incisor and canine, communicates with pyriform and maxillary sinus, passes lateral to nasal bone, through lacrimal groove and into orbit.

Number 4 Cleft

1. *Soft tissue characteristics.* The cleft travels lateral to cupid's bow and medial to the oral commissure, then into lower the eyelid. The ala is rotated superiorly, creating a severe soft tissue deficit between lip and eyelid. The nasolacrimal system and medial canthus are normal (Figs. 11 and 12).
2. *Bony characteristics.* The cleft passes between lateral incisor and canine and then travels lateral to pyriform aperture, through maxillary sinus, medial to infraorbital foramen, and through inferior orbital rim.

Number 5 Cleft

1. *Soft tissue characteristics.* The cleft travels just medial to oral commissure, lateral to the ala and into lower eyelid. There is a short nose and a superiorly rotated alar base (Figs. 13 and 14).
2. *Bony characteristics.* The cleft passes between the premolars, lateral to infraorbital nerve and maxillary sinus, into lateral orbital rim and floor. The maxillary sinus is hypoplastic.

Number 6 Cleft

1. *Soft tissue characteristics.* This facial cleft is similar to that found in Treacher–Collins syndrome. The cheek is furrowed from the oral commissure to lateral eyelid, causing an inferiorly displaced lateral palpebral fissure and lateral colobomas. Ear deformities and hearing deficits are common (Figs. 15 and 16A,B).
2. *Bony characteristics.* There is no alveolar cleft. The occlusal plane is raised on the clefted side. The cleft travels through the zygomatico-maxillary suture and into lateral orbital rim. The zygoma is hypoplastic.

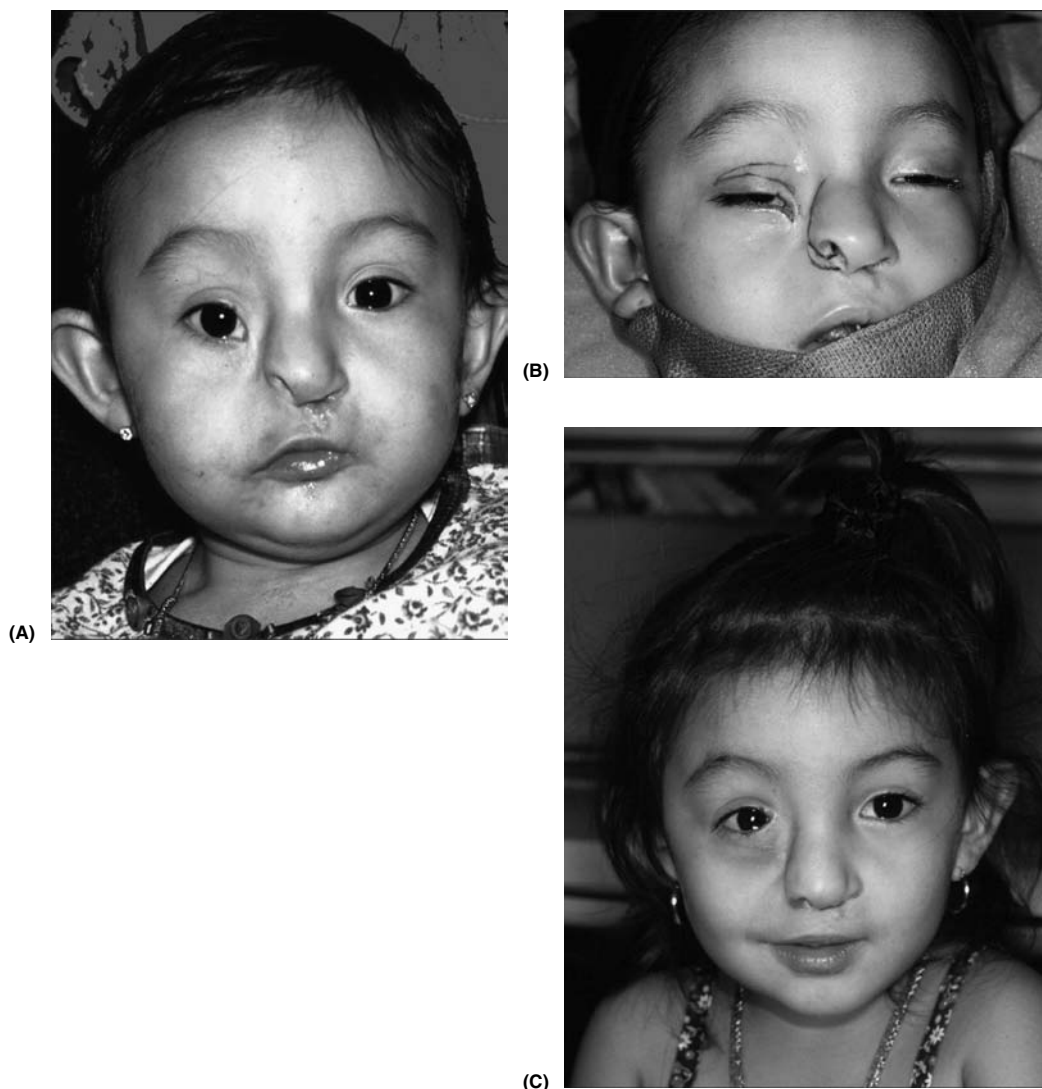


FIGURE 10 Tessier number 3 cleft. (A) Inferior displacement of medial canthus. Superior displacement of alar rim. (B) Intraoperative (surgical incisions marked) Upper to lower lid transposition flap, superior rotation of medial canthus, nasal rotation flap, cheek and lip advancement flaps, and nasal vestibule flap. (C) Postoperative. *Source:* Courtesy of Dr. W. Ozaki.

Number 7 Cleft

1. *Soft tissue characteristics.* The cleft is the same that is seen in hemifacial microsomia and Goldenhar syndrome. There are soft tissue furrows and skin tags from oral commissure to preauricular hairline. The external and middle ears are often malformed. There are occasionally abnormalities of parotid gland, cranial nerves 5 and 7, and the temporalis muscles (Figs. 17 and 18A,B).
2. *Bony characteristics.* The cleft passes through the pterygomaxillary junction. There is hypoplasia of the posterior maxilla and mandibular ramus. The occlusal plane is higher on the affected side, causing an open bite. The condyle, coronoid process, and mandibular ramus are hypoplastic. The zygoma, zygomatic arch, cranial base, glenoid fossa, and sphenoid are typically severely malformed (Fig. 19A–D).

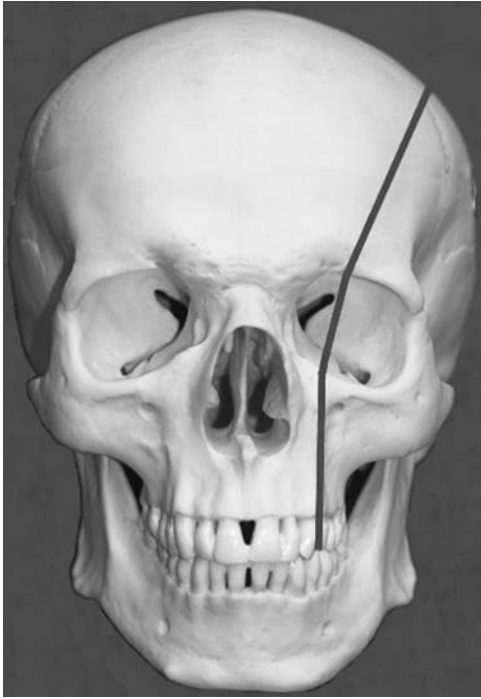


FIGURE 11 Tessier numbers 4/10 craniofacial cleft.

Number 8 Cleft

1. *Soft tissue characteristics.* This is seldom an isolated cleft and more commonly a part of Treacher–Collins syndrome (Figs. 20 and 21). The cleft begins at lateral palpebral fissure and extends onto temporal area. There is a true lateral coloboma and an absent lateral canthus.
2. *Bony characteristics.* There is a cleft of frontozygomatic suture, a hypoplastic zygoma, and downward slanting lateral palpebral fissure.



FIGURE 12 Tessier number 4 cleft. *Source:* Courtesy of Dr. H. K. Kawamoto.

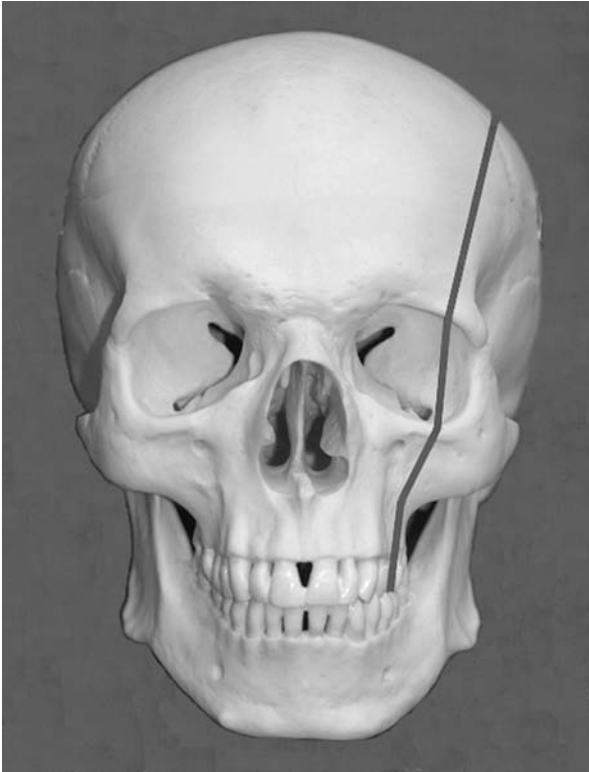


FIGURE 13 Tessier numbers 5/9 craniofacial cleft.



FIGURE 14 Tessier number 5 cleft on the left side. Tessier number 4 cleft on the right side. *Source:* Courtesy of Dr. H. K. Kawamoto.

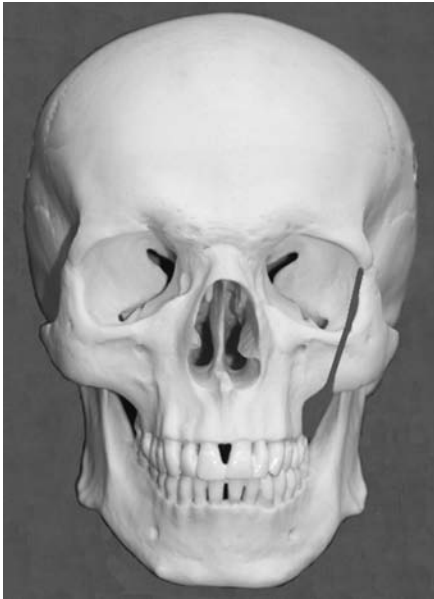


FIGURE 15 Tessier number 6 facial cleft.

Number 9 Cleft

1. *Soft tissue characteristics.* This cleft is extremely rare. The cleft travels through the lateral third of the upper eyelid and eyebrow, distorting the lateral canthus. Cranial nerve seven palsies are common (Figs. 13 and 22A,B).
2. *Bony characteristics.* The cleft travels through the superolateral portion of the orbit, through the greater sphenoid wing and to the squamous temporal bone. Cranial base abnormalities are common.



FIGURE 16 Patient with Treacher–Collins syndrome and bilateral Tessier numbers 6, 7, and 8 facial clefts. (A) Anterior view of cleft. (B) Lateral view of cleft. *Source:* Courtesy of Dr. W. Ozaki.



FIGURE 17 Tessier number 7 facial cleft.

Number 10 Cleft

1. *Soft tissue characteristics.* The cleft begins in the middle of the eyelid and eyebrow, creating colobomas. There is an elongation of the palpebral fissure. Hair tufts often project from the temporoparietal region to the lateral brow (Figs. 11 and 23).
2. *Bony characteristics.* The cleft travels through supraorbital rim, frontal bone, and orbital roof lateral to the supraorbital nerve. Encephaloceles and hypertelorism are common.

Number 11 Cleft

1. *Soft tissue characteristics.* The cleft traverses through the medial eyelid and eyebrow. There is often a downward projecting frontal hair tuft (Figs. 9 and 24).
2. *Bony characteristics.* The cleft is either lateral to the ethmoids and through the supraorbital rim or through ethmoid air cells producing orbital hypertelorism. The cranial base is typically normal.

Number 12 Cleft

1. *Soft tissue characteristics.* The cleft lies medial to the medial canthus, causing lateral displacement of medial canthus. The eyelids are typically normal; however, colobomas may be present. There can be a downward projection of the paramedian frontal hairline (Figs. 7 and 25).
2. *Bony characteristics.* The cleft passes through the frontal process of maxilla and through hypertrophic ethmoid air cells, producing orbital hypertelorism and telecanthus. The frontal and sphenoid sinuses are enlarged, but no encephaloceles are typically present.

Number 13 Cleft

1. *Soft tissue characteristics.* The cleft travels medial to the eyelids, eyebrows, and medial canthus. There is typically a downward projecting, V-shaped, paramedian, frontal hair tuft (Figs. 5 and 26).
2. *Bony characteristics.* The paramedian cleft traverses the frontal bone, courses along the widened olfactory groove and cribriform plate and through the hypertrophic ethmoid air cells. This produces hypertelorism and orbital distopia. Encephaloceles may be present.

Number 14 Cleft

1. *Soft tissue characteristics (widening or duplication).* There is often a frontonasal encephalocele, lateral displacement of the orbits, hypertelorism, telecanthus, and midline hair projections (Figs. 3 and 27).
2. *Bony characteristics (widening or duplication).* There is a bifid crista galli and perpendicular plate of ethmoid and ethmoid pneumatization. The greater and lesser sphenoid wings are rotated causing a short middle cranial fossa.

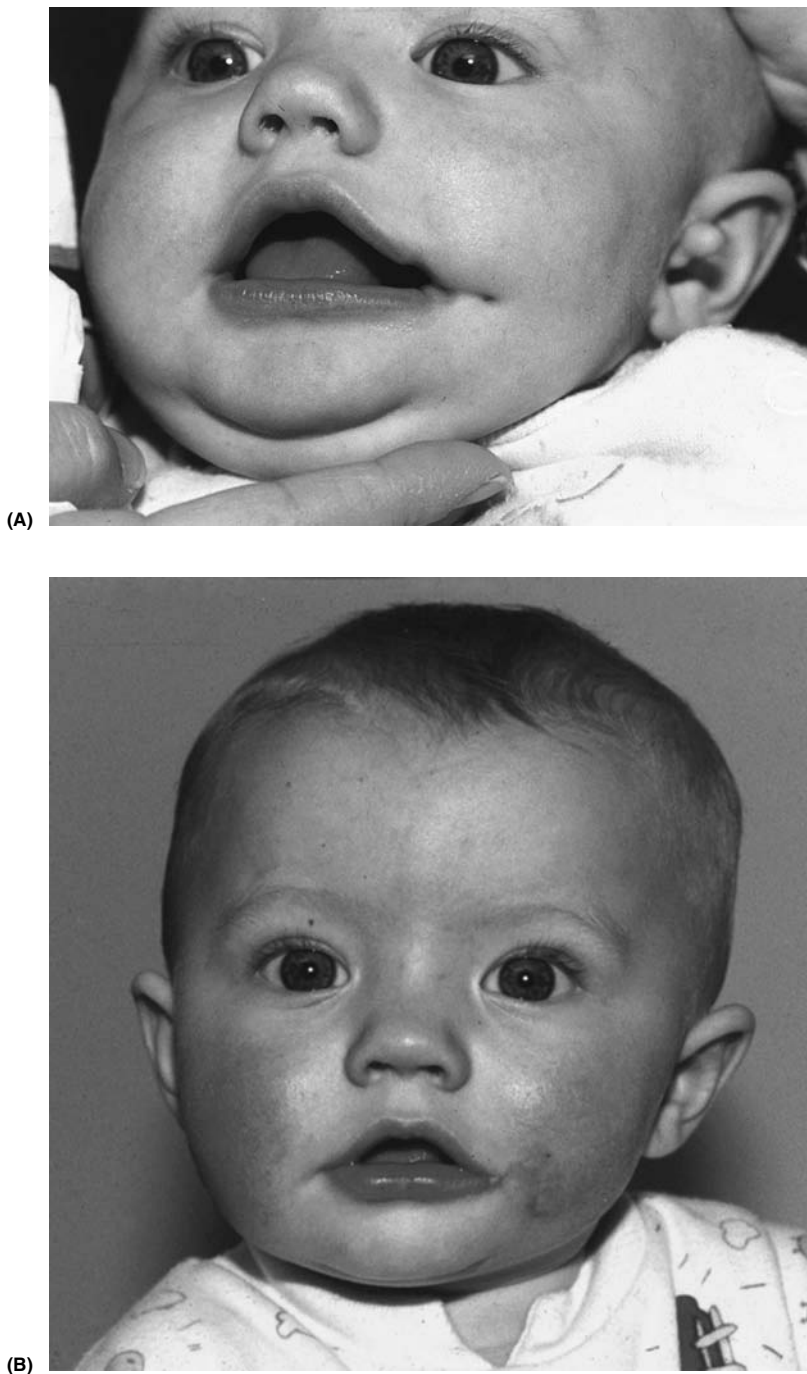


FIGURE 18 Patient with soft tissue number 7 facial cleft. (A) Anterior view of cleft. (B) Postsurgical repair of number 7 facial cleft. *Source:* Courtesy of Dr. H. K. Kawamoto.

3. *Soft tissue characteristics (agenesis or hypoplasia).* Hypoplasia produces holoprosencephalic disorders, microcephaly, hypotelorism, and forebrain malformations (Fig. 28).
4. *Bony characteristics (agenesis or hypoplasia).* There is an absence of midline cranial base, flattened frontal bone, and no pneumatization of frontal sinuses.



FIGURE 19 Patient with craniofacial microsomia and number 7 facial cleft. **(A)** Anterior view of cleft. **(B)** Lateral view of cleft. **(C)** Postsurgical anterior view after mandibular distraction osteogenesis. **(D)** Postsurgical lateral view after mandibular distraction osteogenesis. *Source:* Courtesy of Dr. W. Ozaki.

TREATMENT

1. Treatment plans cannot be rigidly standardized.
2. Timing of surgical intervention is governed by severity of malformation. Surgery is often delayed if the malformation is mild. Early intervention is often mandated if the deformity is severe or there are functional issues.



FIGURE 20 Tessier number 8 facial cleft.

3. Soft tissue clefts and cranial defects corrected during infancy. The midface bone grafting and reconstruction begins after the age of six years. Orthognathic surgery is performed, if necessary, as an adult.
4. Soft tissue clefts are closed with Z-plasties and local tissue flaps. Whenever possible the incisions are placed along aesthetic lines.
5. *Lip reconstruction.* The incisions should be along the philtral column. The lip–vermillion junction should be aligned as in the common cleft lip repair. Muscle continuity should be restored if possible.



FIGURE 21 Patient with Goldenhar syndrome and left Tessier number 8 cleft. *Source:* Courtesy of Dr. H. K. Kawamoto.

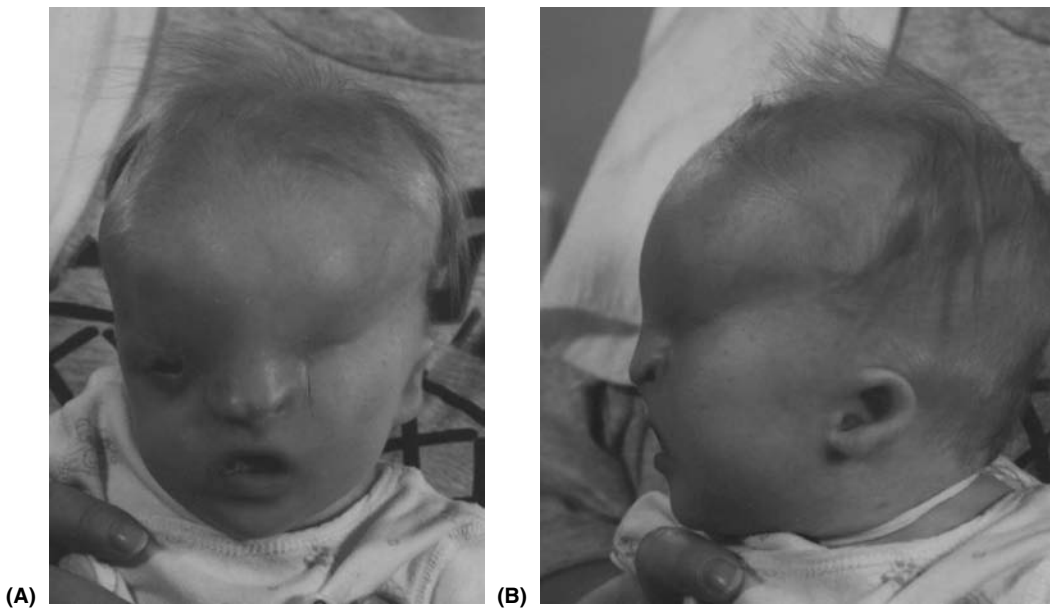


FIGURE 22 Tessier number 9 cleft. (A) Anterior view Tessier number 9 upper facial cleft and number 3 nasal cleft. (B) Lateral view of cleft. *Source:* Courtesy of Dr. H. K. Kawamoto.

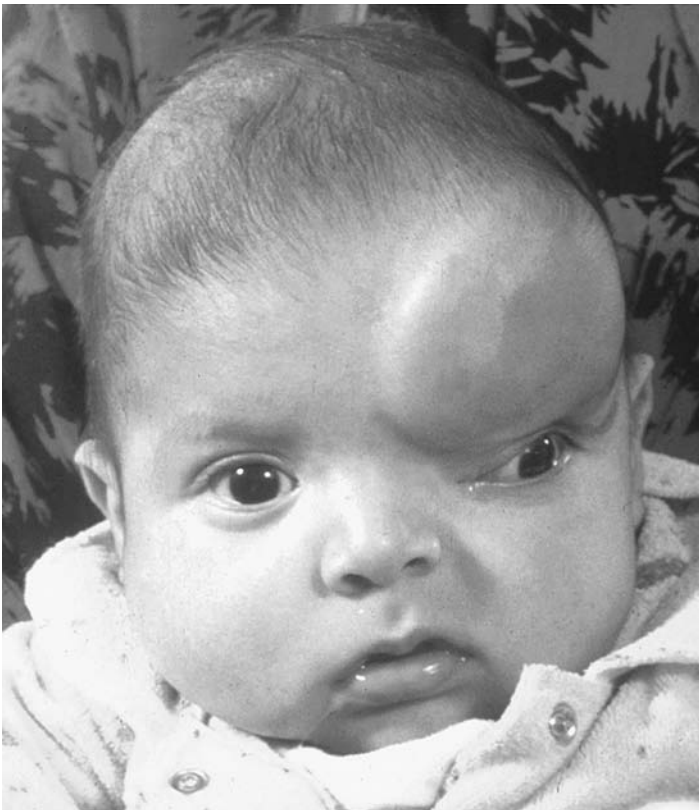


FIGURE 23 Tessier number 10 cleft involving the middle third of the orbit, eyelid, and eyebrow. The orbit is displaced laterally and inferiorly. *Source:* Courtesy of Dr. H. K. Kawamoto.



FIGURE 24 Tessier number 11 cleft. Right number 11 upper lid cleft with loss of medial eyelashes. Right number 3 medial canthus and nasal cleft. *Source:* Courtesy of Dr. B. Novark.



FIGURE 25 Tessier number 12 cleft. Hypertelorism, telecanthus, and paramedian hair tuft. *Source:* Courtesy of Dr. H. K. Kawamoto.



FIGURE 26 Tessier number 13 cleft. Hypertelorism and telecanthus. *Source:* Courtesy of Dr. B. Branter.



FIGURE 27 Tessier number 14 cleft. Widened midline forehead and nasal cleft. *Source:* Courtesy of Dr. H. K. Kawamoto.



FIGURE 28 Tessier number 14 cleft. Narrowed midline forehead and upper lip cleft. *Source:* Courtesy of Dr. H. K. Kawamoto.

6. *Commissure reconstruction.* The commissure cleft should be repaired in a straight-line laterally and a Z-plasty medially. If possible scars should be placed along the nasolabial fold (Fig. 18A,B).
7. *Nasal reconstruction.* The cleft and all surrounding abnormal tissue should be excised. If possible the scars should be placed along aesthetic lines. The cartilage should be repaired and restored to a normal configuration. Cartilage grafts or composite grafts are used if deficiencies persist. Local rotation flaps and Z-plasties are employed for retracted ala. Internal nasal lining can be obtained from the septum. Dorsal nasal projection can be augmented using cantilevered cranial bone grafts (Fig. 10A–C).
8. *Orbital reconstruction.* The orbit should be reconstructed with cranial bone grafts to restore continuity and correct orbital distopia.
9. *Eyelid reconstruction.* The medial canthus must be accurately positioned and secured with transnasal wires. Lateral canthoplasties are performed as necessary and should be positioned several millimeters above the medial canthus. Transposition flaps are used for eyelid skin deficiencies and palatal grafts for conjunctiva deficiencies. Cleft excision and Z-plasties are used to correct number 8 soft tissue clefts.
10. *Eye reconstruction.* Urgent surgical intervention is necessary if the eye is exposed to prevent corneal ulcerations. For anophthalmia, the eyelid and orbit can be enlarged with tissue expanders.
11. *Lacrimal apparatus reconstruction.* When a cleft disrupts the canalicular system it is repaired and then maintained using silastic stents. A dacryocystorhinostomy may be necessary if the lacrimal sac is blocked or absent.

12. Severe hypotelorism is corrected with an intracranial and extracranial facial bipartition.
13. The forehead and orbital are reconstructed with cranial bone grafts if available.
14. If cranial bone is not available rib grafts, iliac crest bone grafts, or alloplastic materials can be used.
15. Alloplastic materials that have been used include: polymethyl methacrylate, hydroxyapatite, resorbable tricalcium phosphate, silicon, and hard tissue replacement (Walter Lorenz[®]; Biomet Microfixation Jacksonville, Florida, U.S.A.).
16. Many craniofacial surgeons will not use alloplastic materials. For those who do, relative contraindications to alloplasts include: placing the material over sinuses, history of craniofacial infection, and inadequate soft tissue coverage.
17. *Lower lip reconstruction.* The cleft should be wedge excised, the vermilion aligned, and the muscle repaired. Z-plasties are used to release the frenulum. The bifid tongue can be wedge excised and closed in layers.
18. *Midline mandible reconstruction.* Mandibular midline clefts should be reconstructed when the child is older, using bone graft as necessary.
19. *Lateral mandible reconstruction.* Severe mandibular deficiencies (absent ramus and condyle) should be corrected with costochondral grafts at the age of six years. Moderate deficiencies can be corrected with distraction osteogenesis, also at approximately six years of age (Fig. 19A–D). Mildly deficient mandibles should not be corrected until the child becomes an adult. For very mild deformities, no correction is needed. For noticeable deformities, orthognathic surgery (maxillary Le Fort I osteotomy, mandibular sagittal split osteotomy and an asymmetric genioplasty) is indicated.
20. The final correction of large soft tissue deficits is done with fasciocutaneous free flaps.

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13 Orthognathic Surgery

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INTRODUCTION

Although it is difficult to estimate, the prevalence of patients with severe skeletal malocclusions that would require orthognathic surgery is significant. In 1990 Proffit et al. estimated that the current number of people in the United States who may benefit from orthognathic surgery is more than 1.5 million (1,2). It is thus important for the plastic surgeon to have a thorough understanding of the diagnosis and treatment of dentofacial anomalies.

Some of the indications for orthognathic surgery in patients with dentofacial deformities include difficulty with proper mastication, inadequate access to oral hygiene, and temporomandibular joint disorders. However, what could initially bring the patient into the surgeon's office may be the unaesthetic appearance of a dentofacial deformity. There are many patients seeking cosmetic improvements in their facial appearance who are truly orthognathic patients with significant dentoskeletal anomalies. Effective communication between patient and clinician is important. It is also important for the plastic surgeon to understand the cosmetic implications of dentoskeletal anomalies. Objective findings, which may be more important to the clinician, and subjective findings, which may be more important to the patient, need to be compared and reconciled in order to achieve optimal results (3).

Although most plastic surgeons get limited exposure to orthognathic surgery, it is imperative that they develop an appreciation for the facial patterns of dentofacial deformities. The evaluation of patients with these deformities might require, in addition to a detailed physical examination, radiographic and dental model analyses. Mastering these diagnostic modalities will help the plastic surgeon better diagnose these patients. The plastic surgeon who goes on to craniofacial surgery, however, is not only expected to be able to diagnose but also to be comfortable with the surgical treatment of these problems. Although dental training is extremely helpful for surgeons who do this kind of surgery, even surgeons with limited dental training can become proficient in orthognathic surgery once they learn certain imperative concepts about dental anatomy and occlusion.

This chapter is an overview of orthognathic surgery for craniofacial surgeons who might have limited dental experience and training. It is clinically oriented and offers a systematic process for the evaluation and treatment of the patient with dentofacial deformities. The chapter will discuss the clinical evaluation and surgical treatment of these patients, and also address interdisciplinary issues such as orthodontic strategies and other nonclinical issues such as model surgery and splint fabrication.

EVALUATION OF THE FACE

The plastic surgeon approaching the patient with complaints related to the face must not only evaluate the soft tissue but also assess the facial skeleton. To this end, the patient must be evaluated with the head in proper position. The patient should be sitting upright, with the examiner sitting directly opposite, and the pupillary and Frankfort horizontal planes should be parallel to the floor. The Frankfort horizontal plane can be estimated by drawing an imaginary line from the tragus to the bony infraorbital rim. In a patient with orbital dystopia, the ears may be used to establish the plane instead of the pupils. Many patients with dentofacial deformities often assume alternative head positions to either improve function or aesthetics (4,5). These compensations must be eliminated in order to properly evaluate the patient. Failure to accomplish this might result in a misdiagnosis and suboptimal treatment.

Ideal facial proportions are useful in determining facial abnormalities. Traditionally, the face may be divided into three equal vertical units separated by horizontal lines at the hairline (trichion), glabella, subnasale, and menton provide landmarks to divide the face into upper, middle, and lower thirds. The lower third is often further subdivided by the stomion, with the upper half being one-third of the lower third and the lower half being two-thirds of the lower third (Fig. 1). As you will see below, changes in lip position, labiomental groove depth, and vertical dimension of the facial thirds can be a key indicator of facial disproportion that may be corrected by orthognathic surgery. In addition, the face may be divided into five equal transverse units (Fig. 2). One should keep in mind that what might have been considered "ideal" proportions can change over the years and certainly differs with race and culture.

In addition to facial proportions, attention should be paid to facial symmetry. If an asymmetry exists, it is important for the surgeon to assess whether the asymmetry is global or confined to a specific segment. Asymmetry confined to the lower third, for example, might indicate a mandibular problem; asymmetry of the lower two-thirds, which might be accompanied by an occlusal cant, could indicate a combined maxillary and mandibular anomaly. When assessing symmetry, it is important to pay attention to the dental midline of the maxillary and mandibular arches. The dental midlines should be aligned with the facial midline as well as with each other (6). Any discrepancy in facial symmetry or dental midline should be recorded carefully.

The relationship between the upper lip and the maxillary anterior teeth is also extremely important. This should be assessed not only at rest but also after having the patient smile broadly. At rest, the upper lip should cover the maxillary incisors except for 2 mm. However, in women, up to 4 mm might be desirable. With the patient at full smile, the entire maxillary incisors should show with minimal or no maxillary gingival show (7). All these relationships apply taking for granted that the maxillary incisors are positioned in their proper inclination on the maxillary alveolar bone as will be discussed in the section on cephalometric tracings. Other variable include the length of the upper lip and the size of the maxillary incisor.

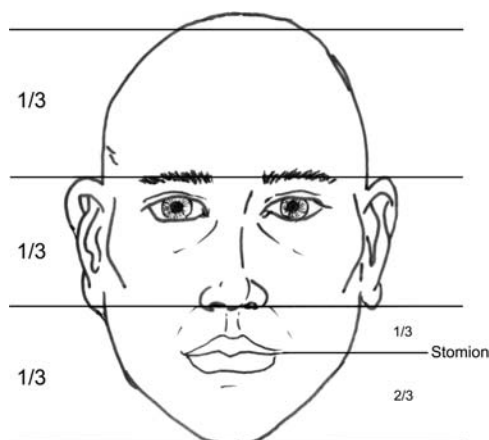


FIGURE 1 Vertical proportions of the face into equal thirds. The lower third is divided into an upper one-third and a lower two-thirds by the point separating the two lips: stomion.

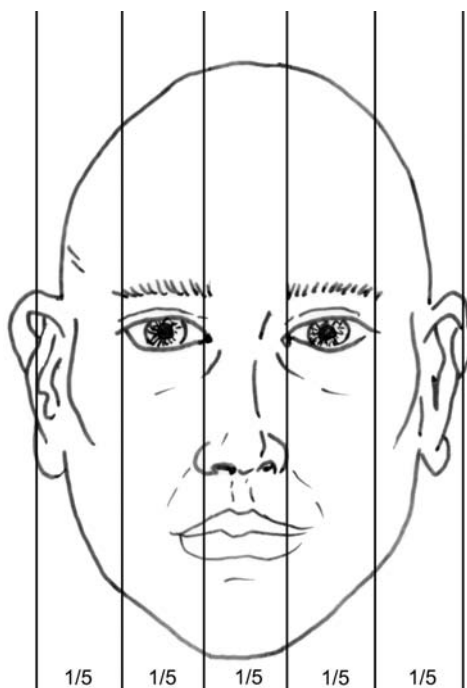


FIGURE 2 Transverse proportions of the face into equal fifths.

When evaluating the face in profile, certain relationships also apply (6–8). In relating the mid-face to the upper face, the Gonzalez-Ulloa zero meridian is of great help (9). This is a line drawn through nasion and at 90° to the Frankfort horizontal line. In the average face, subnasale should fall within this line and the angle between the Frankfort horizontal and the zero meridian should fall from 85° to 92° . This zero meridian can also be used to assess chin position, as the pogonion—the most prominent point on the chin—should lie approximately in a line through subnasale making a 10° angle with the zero meridian (10). In men, the chin should fall on this line or a couple of millimeters anterior, whereas in women, it should be on the line or a couple of millimeters posterior. Once the external facial proportions and relationships are evaluated and recorded, the next step is an assessment of the occlusion.

OCCLUSAL EVALUATION

Any plastic surgeon needs to have a cursory understanding of occlusion in order to evaluate the face. In the 1890s, orthodontist Edward Angle developed the now universally accepted system for describing dental occlusion (Fig. 3) (11). The Angle classification uses the maxillary first molars as the point of reference to describe the anterior–posterior (or mesial–distal) relationship between the maxillary and mandibular arches. In *class I occlusion*, the mesiobuccal cusp of the maxillary first molar articulates within the buccal groove of the mandibular first molar. In this occlusal pattern, the maxilla and mandible are in their normal relationship to each other. In *class II malocclusion*, the mandibular arch is in a posterior position relative to the maxillary arch. Thus, the mesiobuccal cusp of the maxillary first molar articulates anterior to the buccal groove of the mandibular first molar. Class II malocclusion can be further subdivided into divisions 1 or 2, in which the maxillary lateral incisors are flared labially or palatally, respectively. In *class III malocclusion*, the mandibular dentition is positioned anteriorly in relation to the maxillary dentition. Therefore, the mesiobuccal cusp of the maxillary first molar articulates posterior to the buccal groove of the mandibular first molar. Thus, a patient with mandibular prognathism has a class III malocclusion whereas a patient with mandibular retrognathia has a class II malocclusion (12). Angle's occlusal classification system gives the clinician clues to skeletal abnormalities but the clinician must perform a complete physical and radiographic evaluation prior to determine whether skeletal pathology is in the maxilla, mandible, or both.

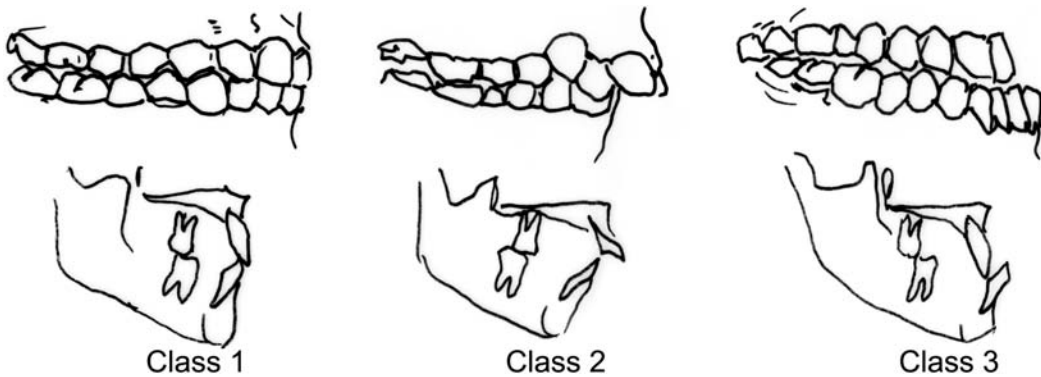


FIGURE 3 Different types of occlusal patterns as described by Dr. Angle.

Understanding the definition of centric relation and centric occlusion is also extremely important for the surgeon who performs orthognathic surgery, as all the model surgery and surgical planning is done from the perspective of centric relation rather than centric occlusion. The two definitions are as follows:

- *Centric occlusion*: Refers to the position of the teeth when there is maximum interdigitation between the upper and lower teeth.
- *Centric relation*: The occlusal relation between the upper and lower teeth when the condyle is seated in its most superior and posterior position in the glenoid fossa; this is a skeletal position which might or might not coincide with centric occlusion.

RADIOGRAPHIC EVALUATION

When the clinical evaluation of the face is indicative of a skeletal problem, further diagnostic studies must be done to confirm the diagnosis. Even though a clinical evaluation can give the relationship between the maxilla and the mandible, it does not assess the relationship of each jaw to the rest of the face. One study that is helpful in diagnosis and essential for treatment planning of orthognathic cases is the cephalometric analysis. This radiographic evaluation will not only assess the relationship between the maxilla and the mandible, but will also assess the relationship of each jaw independently to the cranial base. It is also helpful in assessing the relationship of the dentition to the basal bone of each jaw, which orthodontists can use in planning the movement of teeth within the dental arch.

The cephalometric analysis is done by taking a lateral and frontal skull view and evaluating certain skeletal points and their relative relationship. This is a way of relating the position of the maxilla and mandible independently to the cranial base. The measurements and angles obtained during the cephalometric tracing are compared with certain established “normal” ranges. Some important landmarks (Fig. 4) on the lateral cephalogram that help with this task include:

- *Sella* is the center of the pituitary fossa
- *Nasion* is the most anterior point at the nasofrontal junction
- *A-point* is the deepest midpoint on the maxillary alveolar process between the anterior nasal spine and the alveolar ridge
- *B-point* is the deepest midpoint on the mandibular alveolar process between the crest of the ridge and pogonion
- *Gonion* is the most inferoposterior point at the angle of the mandible *Gnathion* is the cephalometric intersection of facial and mandibular planes
- *Pogonion* is the most anterior point along the contour of the symphysis
- *Menton* is the lowest point on the contour of the mandibular symphysis

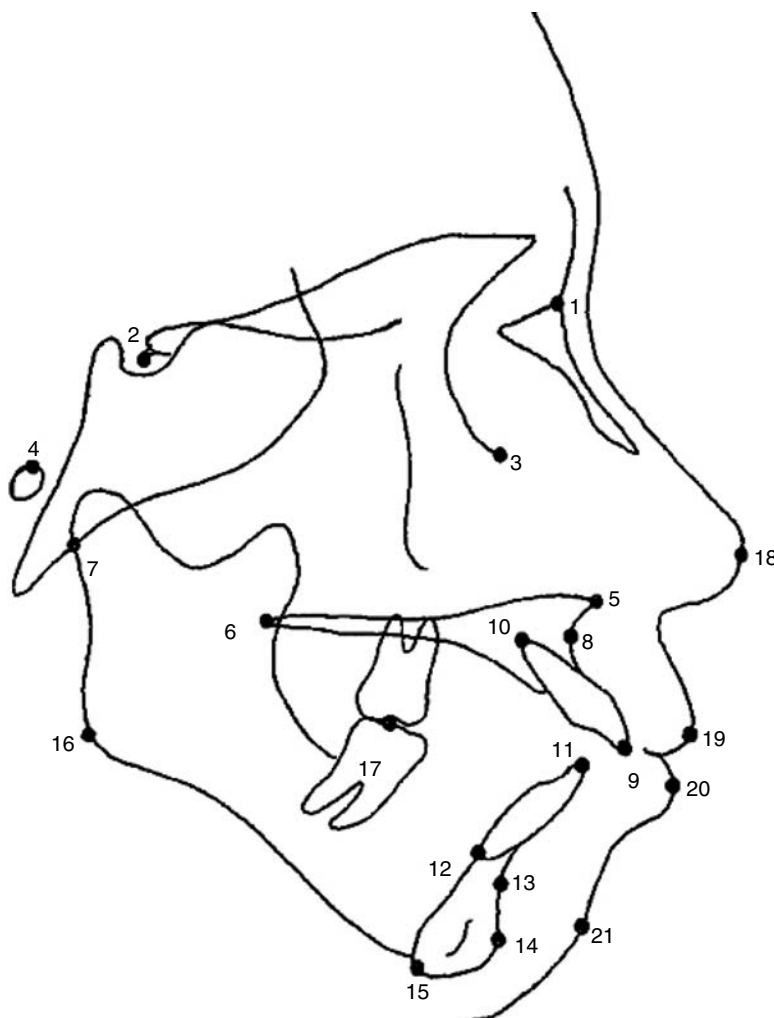


FIGURE 4 Landmarks used: 1, nasion; 2, sella; 3, orbitale; 4, porion; 5, anterior nasal spine; 6, posterior nasal spine; 7, articulare; 8, A-point; 9, incisal end of maxillary incisor; 10, apex of maxillary incisor; 11, incisal end of mandibular incisor; 12, apex of mandibular incisor; 13, B-point; 14, pogonion; 15, menton; 16, gonion; 17, articulation of maxillary and mandibular molars; 18, pronasale; 19, upper lip; 20, lower lip; 21, soft tissue pogonion.

- *Orbitale* is the lowest point on the inferior bony border of the left orbital cavity
- *Porion* is the most superior extent of the external auditory meatus
- *Mandibular plane* is formed by the line joining gonion with menton
- *Frankfort horizontal plane* is formed by the line joining porion with orbitale
- *Aesthetic line* is formed by the line joining the tip of the nose with the chin

By utilizing any of the cephalometric analyses available, the clinician can gain useful information as to the relationship of the maxilla and mandible to the cranial base as well as to each other. This information is useful in deciding whether the skeletal abnormality is confined to only one jaw or whether it includes both the maxilla and the mandible. Although most of the cephalometric analyses are somewhat complex and assess multiple parameters of the facial skeleton, the orthognathic surgeon needs to be familiar with certain angles and relationships that are crucial to the treatment planning of any orthognathic case. Some of the more pertinent parameters are as follows (Fig. 5):

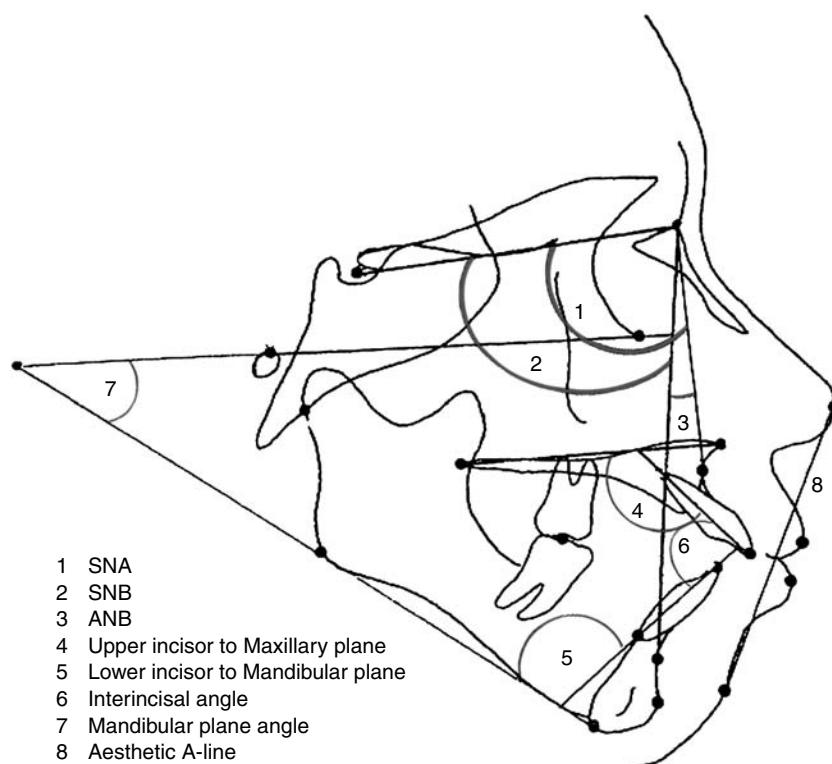


FIGURE 5 Cephalometric tracing. See text for further description.

- SNA is the angle that relates the maxilla to the cranial base (mean: $82 \pm 3^\circ$).
- SNB is the angle that relates the mandible to the cranial base (mean: $80 \pm 3^\circ$).
- ANB is the angle that relates the jaw position relative to one another. The angle should always be zero or positive to 2 mm.
- *Upper incisor to maxillary plane*: This angle gives you an indication of the inclination of the maxillary incisors relative to the maxillary alveolar bone. An abnormal incisal plane will affect the relationship of the upper lip to the maxillary incisors and might impact the clinical assessment of the vertical positioning of the maxilla. For example, abnormally flared maxillary incisors will mask true incisal show and this, in turn, can lead to underestimation of vertical maxillary excess (VME).
- *APo plane* is a line from A-point to pogonion and becomes a line of reference for the protrusion or retrusion of anterior teeth.
- *Lower incisor to mandibular plane* is the lower incisor measured from the denture plane (APo) but is planned from the APo relationship produced with orthodontic treatment. The ultimate height of the occlusion is keyed from the lower anterior area.
- *Mandibular plane angle* relates the posterior facial height to the anterior facial height. It is derived from the angle between the Frankfort horizontal plane and the mandibular plane. The mean mandibular plane angle is $21 \pm 3^\circ$ and is more obtuse in patients with an anterior open bite and/or micro- or retrognathia. Patients with a deep bite and/or short face syndrome tend to have a more acute mandibular plane angle. Patients with abnormally acute mandibular plane angles present a more difficult treatment challenge, as increasing this angle involves lengthening the muscular pterygo-masseteric sling, an unstable maneuver that is prone to relapse.
- *Aesthetic line (A-line)*: The nose tip point is called pronasali. The most prominent chin point is labeled propogonion. The A-line serves as a reference for the protrusion of the lips. The lower lip is a reference to which the upper is compared.

OTHER DIAGNOSTIC TESTS

If the clinical and cephalometric evaluation point to a skeletal abnormality, then orthognathic surgery should be considered. Further information might be obtained from diagnostic dental models, which can help to assess the inclination of the teeth, the transverse dimensions, the total arch length, and the current occlusion. Models taken before the surgery will help in deciding the way the teeth will fit during and after orthognathic surgery. At this point, orthodontic consultation is needed as most orthognathic cases require some degree of orthodontic therapy.

ORTHODONTICS

The orthognathic surgery patient will usually require orthodontic treatment prior to surgery. Orthodontic preparation is based on a thorough and systematic diagnosis and treatment plan prepared jointly by the orthodontist and the surgeon. This serves to identify the patient's problems and to develop a treatment plan that will most efficiently and effectively address the patient's concerns. Clinicians should listen carefully to the patients' concerns and elicit their perceptions of orthognathic surgery. Although many patients understand the relationship between aesthetics and facial skeletal relationships, others are unaware that the aesthetic concerns originate from problems with their jaws and teeth (13).

In most malocclusions, there is a certain degree of skeletal-dental compensation of the dentoalveolar component. All these compensations must be identified and reversed by the orthodontist prior to orthognathic surgery, in order to achieve a stable postoperative result. Orthodontic goals prior to orthognathic surgery include removal of all dental compensations and achievement of proper tooth alignment in the arch and in relationship to the alveolar bone. The final orthodontic goal is to position the teeth ideally in the context of the planned surgical procedure and facial aesthetics. The success of the surgical correction will be determined by the orthodontist's ability to move the teeth into the predetermined and planned positions. It should be the orthodontist's goal to position the teeth in an ideal arrangement within the basal bone, and allow for the orthognathic surgery to position that basal bone within the proper anatomical relationship to the cranial base and between maxilla and mandible. This will in turn result in ideal and stable skeletal, occlusal, and neuromuscular relationship and in a physiological temporomandibular joint relationship to the cranium.

The orthodontic treatment associated with orthognathic surgery is divided into two distinct phases: pre- and postsurgical. Presurgical orthodontics, described above, can last from 12 months to 2 years. Postsurgical orthodontics begins approximately four to six weeks after surgery, and has the main goal of achieving the optimal and most stable occlusal relationship.

At the completion of the postsurgical orthodontic phase, there should be an optimal occlusion. Retainers should then be used to ensure the stability of the orthodontically moved teeth until the periodontal supporting mechanism has reorganized in the new positions. Required retention time varies widely and needs to be evaluated for each individual case. This phase of treatment may range from 6 to 12 months (13–15).

PLANNING THE SURGICAL MOVEMENT

Once the orthodontist, in consultation with the surgeon, determines that the presurgical orthodontic phase is complete and the patient is ready for surgery, a complete new set of records must be obtained, as the orthodontic treatment is likely to have changed tooth angulation and position, which will impact the surgical treatment plan. A clinical examination, cephalometric evaluation, and diagnostic models must be completed, and it is with this new set of data that the surgery should be planned. This new diagnostic information should be obtained within two weeks of the surgical date, as to prevent ongoing orthodontic changes from having a significant impact on the surgical plan.

The vertical position of the maxilla should be the first consideration in deciding which surgical movements to make. The anterior maxillary position is arrived at clinically by examining the incisal show at rest. As previously stated, 2 mm is ideal in males whereas up to 4 mm

might be desirable in females. The posterior maxillary movement should be adjusted to obtain adequate occlusion. For example, in patients who present with an anterior open bite, the posterior maxilla will require more impaction than the anterior maxilla in order to close the open bite.

The anteroposterior (AP) movements of the maxilla are usually best determined by the cephalometric study. Taking clinical assessment into consideration whenever possible, one should place the maxilla in order to achieve a normal position to the cranial base by utilizing the cephalometric tracing. This position is determined by an SNA angle that is within the accepted norm. The transverse position of the maxilla is best determined in the diagnostic models, which will easily allow the surgeon to determine any potential cross-bites. If the transverse dimension of the maxilla needs to be modified, then segmental maxillary surgery should be performed.

Once the maxillary movements are treatment planned, consideration should be given to the new position of the mandible in reference to the new position of the maxilla. If the new mandibular position does not result in an optimal occlusion and position, then mandibular surgery must be performed to accomplish this end. It is important to appreciate that any time the vertical position of the maxilla is altered, the mandible will achieve a new position by rotating (autorotation) along the axis created by the temporomandibular joint. If the maxilla is impacted, the mandible will rotate in a counterclockwise direction, which will have the net result of placing the mandible more anterior in relation to the cranial base. The opposite will be true if the maxillary movement is in an inferior direction.

MODEL SURGERY

After the new set of diagnostic information is obtained and before the patient is taken to the operating room, the final surgery is planned and performed as model surgery.

The purpose of the model surgery is to assist the surgeon in transferring three-dimensional movements directly to the patient during orthognathic surgery. It also allows the surgeon to confirm the surgical movements and provide quantitative data that are useful intraoperatively (16). This is achieved by obtaining specific measurements and reference points from dental casts, and preparing custom-made surgical splints. These splints are then used to guide the movements of the osteotomized segments before performing rigid fixation.

Properly taken and poured dental models are essential in the model surgery process, and so is the way in which these models are mounted. Ideally, all models should be mounted on a semiadjustable articulator after a facebow transfer, which relates the maxilla as it is positioned three-dimensionally on the face utilizing the Frankfort plane as the horizontal axis. The mandible is mounted on the articulator utilizing a bite registration from the patient, while he or she was in centric relation—the occlusion when the condyles are positioned in their most superior and posterior position in the glenoid fossa. This is done by placing gentle pressure symmetrically on the chin in a posterosuperior direction, while the patient bites into a wax wafer. The reason centric relation is utilized for the mounting (as opposed to centric occlusion, the point of maximal dental interdigitation) is that centric relation is more easily reproducible in the operating room by manipulating the mandibular condyles into that position intraoperatively.

Facebow transfers and semiadjustable articulators are ideal in all cases and are highly recommended in patients with anticipated two-jaw procedures, temporomandibular joint dysfunction or facial asymmetries (17). For practical purposes, however, model surgery for patients with anticipated single-jaw procedures with no other comorbid factors can be mounted in simple, hand-mounted hinge articulators. In this instance, maxillary and mandibular casts are held together in the ideal centric occlusal relationship with sticky wax and then mounted on a simple hinge articulator. Once mounted, the final surgical splint is fabricated.

Once the casts are mounted on the articulator, the model surgery can begin. Certain principles must be followed in performing model surgery in order to ensure accurate splints and optimal surgical results (17–19). Traditionally, for orthognathic surgery involving the maxilla, dental casts are mounted using a facebow transfer to relate the maxilla to the cranial base and an occlusal bite registration in centric relation for the mandible. Reference lines, both vertical and horizontal, are placed on the maxillary cast in order to measure the movements made.

The horizontal reference marks are usually made at 10 mm intervals from the articulator mounting ring. Dental landmarks are then utilized to make a series of measurements (with a Boley gauge) in order to document the preoperative anatomic position of the maxilla. This is done at several points on the cast—for example, at the anterior midline and at the mesiobuccal cusp of each maxillary first molar.

If the anticipated maxillary surgery involves impaction of the maxilla, it is necessary to remove additional plaster from the mounting to provide clearance. Conversely, stone is added if the maxillary movement is in the inferior position. The cast is then ideally positioned and the incisal guide pin is adjusted so that it is consistent with the planned vertical change in the position of the maxilla. Another way to determine the amount of maxillary movement on the cast is to measure from the mounting plate to the occlusal plane on three different positions on the cast. After the stone is removed or added and the maxillary cast is repositioned, the same measurements are performed until the desired amount of vertical maxillary movement is achieved. The amount of movement in the horizontal plane is measured utilizing the vertical markings on the cast as a reference.

The maxilla is then remounted onto the upper member of the articulator. Vertical, AP, and transverse measurements are repeated in order to confirm the three-dimensional surgical movements of the maxilla. This information is compared with clinical and cephalometric data, and with the proposed treatment plan. With the maxillary and mandibular casts secured in their final ideal position, an acrylic occlusal splint is constructed. The splint is then used to transfer these vertical and horizontal changes in maxillary position intraoperatively. In the operating room, the final vertical position of the maxilla is confirmed using an external reference point (e.g., glabellar pin or temporary tattoo on the medial canthus region) during the actual surgical movement.

For segmental maxillary surgery, the presurgical evaluation of the maxillary cast must include additional measurement at dental landmarks in order to document the surgical movement of each dentoalveolar segment. In this case, the maxillary cast is removed from the upper member of the articulator and divided into two or three segments, depending on the preoperative treatment plan. These segments are then positioned opposite the mandibular case, making sure that the dental midlines are aligned, and the transverse dimension is such that the occlusion achieved is optimal. Sticky wax and mounting plaster are used to secure this newly establish maxillary arch form. The maxillary cast is then returned to the semiaadjustable articulator and positioned anteroposteriorly on the mounted mandibular cast. The remounted maxilla may then be measured to confirm the three-dimensional changes of the segments. An acrylic occlusal splint is then constructed, as mentioned above. When segmental surgery is being performed, however, this splint is secured to the maxillary teeth during the surgery and left in place for four to six weeks. To accomplish this, small perforations are made on the splint at the location of each interdental space. Through this space, a thin orthodontic wire is passed and the splint is thus secured to the arch wire.

For combined maxillary and mandibular surgery, the model surgery must be completed in stages. After the planned maxillary movements are made, the maxillary cast is mounted in its new position and an intermediate splint is fabricated, allowing the mandible to rotate into occlusion on the articulator. After this is accomplished, the mandibular cast is separated from the mounting plate and remounted on the articulator in the optimal occlusion. The final splint is then fabricated with the mandible in this position. It is usually not necessary to take measurements of the degree of mandibular movements, although this can be done by making similar horizontal and vertical markings on the cast. This information can be useful when the movements are at the extremes, and can alert the surgeon to the potential for significant relapse.

OPERATIVE PROCEDURES

Maxillary Deformities

VME is characterized by excessive tooth display when the lips are in repose, excessive gingival exposure on smiling, lip incompetence and, at times, mentalis muscle strain as the patient tries to force the lips closed. This condition is often associated with an open bite. The face height

is always long, and the chin is rotated downward and posteriorly. This condition is exaggerated by the presence of a short upper lip or maxillary protrusion. VME can be seen with class I, II, or III occlusions.

When VME is significant enough to require surgery, it is usually treated with Le Fort I osteotomy and superior repositioning of the maxilla. If the maxillary impaction is significant, and depending on the presurgical occlusion, sometimes the autorotation of the mandible after the maxilla is impacted results in a class III occlusion that needs to be corrected with a mandibular pushback osteotomy.

It is important to note that because the vertical growth of the maxilla is the last vector to cease, the excessive vertical development may continue growing later than expected (20). This, in turn, can result in significant relapse. It is thus recommended that maxillary impaction be performed only after facial growth is complete. When the patient is experiencing significant psychological sequelae due to the skeletal deformity, the operation may be performed before growth is complete. In these situations, it is prudent to advise the patient and parents that secondary surgery might become necessary due to relapse.

Vertical maxillary deficiency (VMD) is often present with other skeletal abnormalities, such as AP or transverse maxillary deficiency or mandibular prognathism. The lower face height is always reduced, and the freeway space is excessive. Often, the maxillary incisors are completely covered by the upper lip at rest, with only a portion of the crowns exposed when smiling, and a proper sized mandible will appear prognathic because of the overclosed position. These patients usually have a class III malocclusion with differences between centric relation and centric occlusion. Treatment of VMD usually involves Le Fort I osteotomy with downgrafting, often in combination with a mandibular osteotomy (21).

Maxillary AP deficiency, a condition typically seen in cleft patients, is characterized by paranasal deficiencies, deficiency of the infraorbital region and lack of zygomatic prominence. A major soft tissue characteristic found in these patients is the upper lip behind the lower lip. These patients have a class III malocclusion with compensatory flaring of the maxillary anterior incisors in the true condition. Cephalometric analysis easily confirms the clinical diagnosis and helps determine whether the mandible contributes to the deformity and the malocclusion. Treatment of this condition involves maxillary advancement. For significant advancements of more than 10 mm, sometimes bone grafting is recommended in order to improve the stability of the maxilla in its new position (22).

Le Fort I Osteotomy

Le Fort I osteotomy, with or without segmentation, can be used to correct a variety of maxillofacial deformities in all planes (Fig. 6). Nasotracheal intubation is performed and the tube is secured to the septum with a 2-0 silk suture. As with all facial surgery, positioning of the patient at the time of surgery is important. The head should be parallel and slightly elevated to the plane of the operating table. In order to measure the movements of the maxilla, especially on a vertical dimension, it is useful to use an external landmark—either a pin placed on the glabella or a tattoo on the medial canthus. Measurements from this point of reference to either the teeth, the orthodontic wires, or the orthodontic brackets in a particular tooth on either side of the midline should be taken preoperatively and recorded. These measurements are then utilized to arrive at the final position of the maxilla after the anticipated movements are made. Some surgeons prefer to make their reference marks and measurements on the maxilla itself, but these marks are not very reliable as sometimes they are lost during the osteotomy. In addition, sometimes there is telescoping of the segments or loss of the alignment of the marks with the maxillary movements.

After local anesthetic infiltration, the intraoral incision is made along the maxillary vestibule from the region of the first molar to the contralateral first molar. A small cuff of freely movable mucosa should be left behind on the maxilla, in order to make closure easier at the end. Once the maxilla is exposed, subperiosteal dissection superior to the incision along the maxillary walls should be carefully performed. The dissection should extend superiorly until the infraorbital nerve is visualized, and posteriorly until the periosteal elevator reaches the pterygoid plates. Medially, the piriform aperture should be visualized and the dissection

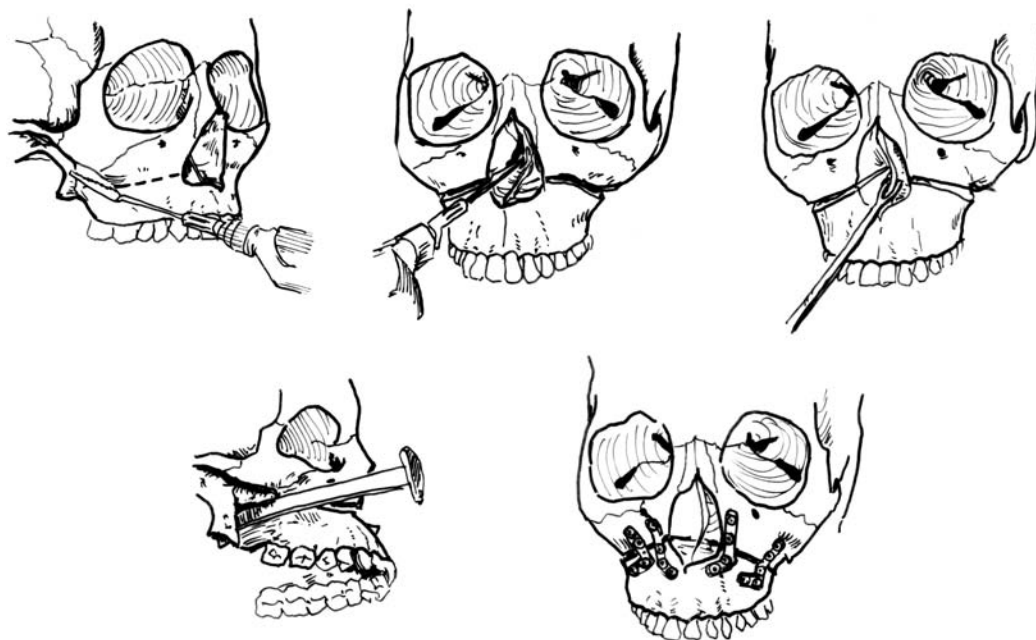


FIGURE 6 The steps in a Le Fort I osteotomy.

should be extended to the nasal cavity until the nasal mucosa is dissected free from the nasal floor and the lateral walls. The dissection is extended superiorly to the inferior turbinate.

This is the time to make reference points. Vertical and horizontal reference marks can be made at the piriform and the zygomatic buttress regions. The design of the lateral maxillary osteotomy is tailored to the aesthetic needs of the patient. For instance, if the patient requires greater augmentation of the malar region, the osteotomy can incorporate the zygomaticomaxillary buttress as well as the higher aspect of the lateral maxillary wall. The osteotomy should always be designed so that it terminates inferiorly in the piriform aperture region (under the inferior turbinate). This minimizes the risk to the nasolacrimal system.

The osteotomy should be designed high enough as to prevent damage to the tooth roots—at least 5 mm away from the apex of the dental roots. On average, the apex of the dental roots is approximately 25 mm from the occlusal plane, although the canine root is usually several millimeters longer. It is usually helpful to utilize a sterile pencil or a marking pen to mark the osteotomy design on the maxilla.

Starting at the zygomaticomaxillary buttress, the reciprocating saw is utilized to start the osteotomy in the anterior direction and end at the floor of the nose in the piriform rim. Once this cut is completed, a similar maneuver is performed in a posterior direction from the zygomaticomaxillary buttress. Either the reciprocating saw or an osteotome can then be utilized to perform the osteotomies of the medial antral wall and the nasal septum and vomer. In performing these osteotomies, it is important to pay particular attention to the location of the endotracheal tube to prevent damage to the tube. To this end, the osteotomies of the medial antral wall should be directed laterally or away from the tube. If the nasal septum and vomer are to be cut with the saw, it should be started on the nostril with the tube and the cut should be made in a direction away from the tube. Finally, curved osteotomes should be utilized to separate the pterygoid plates from the maxilla. This cut should be made in a medial and inferior direction, while the surgeon's finger is placed on the palate in the area of the hamular notch so that the osteotome can be palpated after the cut is completed. Once all the osteotomies are completed, the maxilla will be ready for down-fracturing.

Thumb pressure can be utilized on the anterior maxilla (not the teeth) to down-fracture the maxilla by applying the pressure in an inferior direction. Sometimes counterpressure can be

utilized to help in this step by placing the index finger in the area of the zygoma. If the down-fracture is not easily accomplished with finger pressure, it is important to check all osteotomies to ensure that they are complete. An alternative is to use the Rowe's disimpaction forceps to accomplish the maxillary down-fracture.

As the maxilla is mobilized, a periosteal elevator is utilized to complete the dissection of the nasal and antral mucosa from the maxilla. It is important to keep as much of the mucosa intact as possible, as this will decrease bleeding. Once the maxilla moves freely in an inferior direction, mobilization laterally should be performed. Curved osteotomes are placed one side at a time in the area of the pterygoid-maxillary suture and pressure is applied anterior and toward the opposite side to allow the maxilla to come forward and rotate in the opposite direction. Any remaining posterior bony attachments that prevent this movement are then removed with rongeurs.

When the maxilla is fully down-fractured, bleeding arising from mucosal tears and osteotomy sites can be easily controlled by temporary packing. At times, the descending palatine vessels are easily visualized running vertically in the posterior maxilla near the medial wall of the maxillary sinuses. If these vessels are intact, they should be preserved to augment the blood supply to the maxillary segment. If these vessels are severed, injured, or bleeding, it is acceptable to cauterize or ligate them to control the bleeding. The blood supply to the down-fractured maxilla comes from the soft palate through the bilateral ascending pharyngeal arteries. Thus, sacrificing the descending palatine arteries should in no way compromise the vitality of the maxillary segment, and some surgeons opt to cauterize these vessels routinely as part of their Le Fort osteotomy technique.

Depending on the movements that are to be accomplished, bony interferences are removed with either rongeurs or a bur. Posterior movements of the maxilla are sometimes the most difficult to accomplish, as it is a technical challenge to remove bone in the area of the posterior maxilla and around the pterygoid plates. It is important to trim the nasal septum and vomer when maxillary impaction is planned, as failure to do this can result in septal deviation from a septum that remains too long when the maxilla is impacted. When the maxilla is to be moved anteriorly or inferiorly, consideration should be given to placing bone grafts in the gaps that invariably will be present once these movements are accomplished.

Once all interferences are removed, the anticipated movements are measured against external landmarks. The splint is then placed and maxillomandibular fixation (MMF) is accomplished with the splint in place by utilizing wire or elastics to the surgical hooks on the orthodontic wires. It is important at this stage to utilize the mandible as a point of reference. The maxilla should be able to hinge into the desired position utilizing the mandibular condyles as the axis of rotation. It is important to manipulate the maxilla into position while the condyles are fully seated in the glenoid fossa (centric relation). Arriving at the desired maxillary position should be accomplished with light finger pressure. Any interference present should be removed as they could lead to postoperative relapse. With the maxilla in the proper position, and after all interferences are removed, the next step is to accomplish fixation. In the maxilla, this is usually done with 2 mm plates. Usually two L-shaped plates are utilized per side and are positioned at the zygomaticomaxillary buttress and the piriform rim buttress areas.

After the fixation is completed, the new maxillary position is checked with the external landmarks. It is important to check both sides of the midline to ensure that there is no canting of the maxillary plane. Once the surgeon is satisfied with the new maxillary position, the incision is closed, usually with a 3-0 or 4-0 resorbable suture such as Chromic or Vicryl. For maxillary impaction or anterior movement, some surgeons recommend an alar-cinch stitch. This is a horizontal mattress suture through the alar base bilaterally to prevent alar flaring postoperatively, and it is usually done with a permanent suture such as 2-0 nylon. If the patient presented with a short maxillary lip, a V-Y closure of the maxillary incision can be incorporated to lengthen the lip (Fig. 7) (23).

Segmenting the Maxilla

Sometimes—when there are steps in the occlusion, for example, or when the transverse dimension of the maxilla needs to be changed—the maxilla must be segmented during the

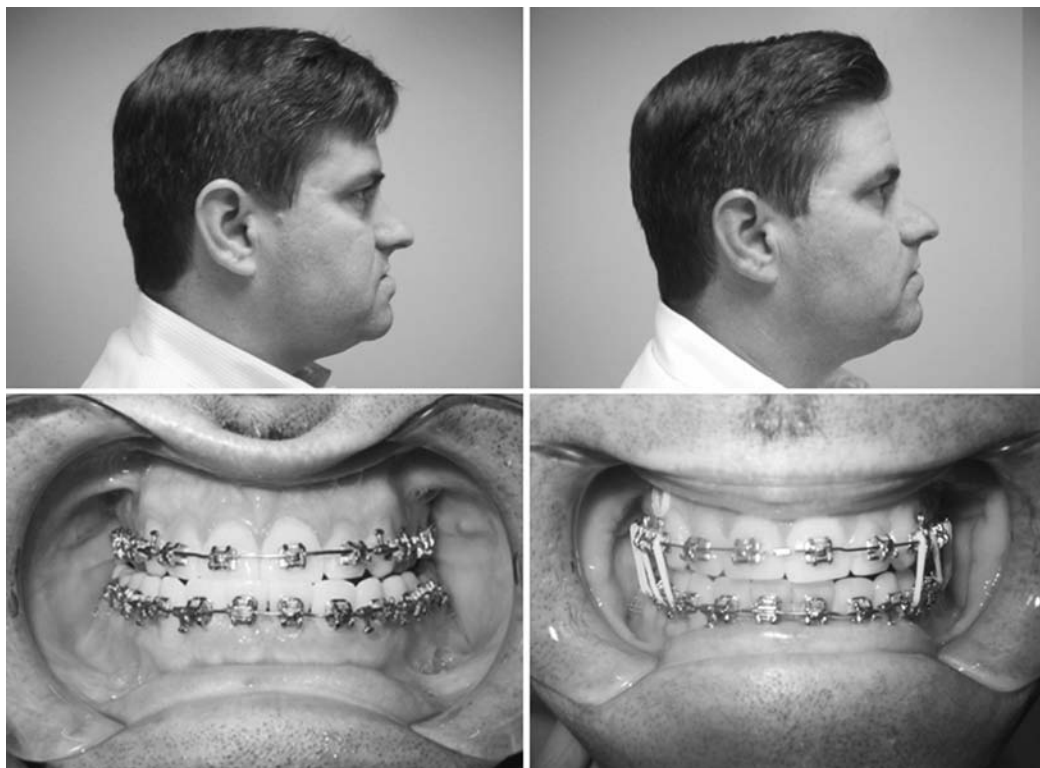


FIGURE 7 Patient with class II malocclusion after a Le Fort I maxillary advancement.

Le Fort I osteotomy. Perhaps the most common segmentation is a paramedian osteotomy that divides the maxilla into two segments and allows for the increase in the transverse dimension of the maxilla. Other patterns of segmentation are possible. When trying to correct bimaxillary protrusion, for example, a transverse osteotomy, which divides the maxilla in an anterior and a posterior segment, could be used. It is also possible to combine the different osteotomies to divide the maxilla into three or more segments.

When segmenting the maxilla, the surgeon must consider the desired movements and take into consideration local anatomical concerns. Sectioning of the midline of the palate is avoided, since the bone is thick and the palatal soft tissues are thin and more likely to rupture when damaged or stretched. The palatal bone is thinner laterally and easier to cut, and the soft tissues laterally are thicker and less likely to rupture when stretched. When significant arch expansion is anticipated, use of two paramedian osteotomies minimizes the bone defects and disperses the soft tissue tension over a broader area.

Segmentation is usually performed after the maxilla is down-fractured. The surgeon places a finger on the palatal mucosa to prevent injury and uses a reciprocating saw to make the osteotomies through the nasal floor. As soon as the surgeon palpates the saw coming through the bone, pressure is released on the saw in order to keep the mucosa intact. In order to complete the osteotomy through the alveolar ridge, the attached mucosa is gently reflected from the bone surrounding the collar of the teeth with a fine chisel. The osteotomy should be carried out in the appropriate interdental space between the tooth roots to prevent damage to same. It is usually not difficult to see the outline of the particular roots through the alveolar bone, as the roots form a slight prominent area on the bone, while the interdental space bone is flatter or even depressed. When it is difficult to ascertain the location of the roots, a panoramic radiograph or periapical dental radiographs can be of great help. Once the osteotomies are complete, mobilization of the different segments is accomplished after the orthodontic wire is cut in the same interdental space where the osteotomy was performed.

After the mobilization is completed, the splint is placed and secured to the different maxillary segments. It is imperative that the different segments fit passively on the splint; therefore all interferences must be removed to accomplish this. In cases where the final position of the segments allow for significant bony gaps between the segments, small bone grafts can be wedged in these gaps in order to stabilize the different segments. Although allogeneic materials can also be used, it is preferable to use autogenous bone.

In cases where segmentation of the maxilla is performed, it is also imperative to leave the final splint attached to the maxilla for a period of four to six weeks. This is accomplished by making small orifices on the splint in the interdental areas and securing the splint to the orthodontic hooks with fine wire.

Mandibular Deformities

In patients who present with the maxilla in the correct position, but with either a class II or class III malocclusion, it is the mandible that should be addressed. Mandibular prognathism and class III malocclusion usually presents with maxillary deficiency, but occasionally it presents as an isolated condition. Oftentimes, the skeletal discrepancy is much worse than the occlusal discrepancy because there is often a significant amount of dental compensation in these patients. Thus, it is important to reassess these patients after the orthodontist removes all dental compensations. It is then that the true degree of skeletal discrepancy will truly become apparent.

The procedure of choice for treatment of mandibular prognathism is a sagittal split osteotomy. Although some surgeons advocate an intraoral vertical ramus osteotomy (IVRO), especially for large posterior movement and when there is a need for an asymmetric setback, this procedure has fallen out of favor, because it requires intermaxillary fixation. As previously discussed, surgery should be undertaken only after dental compensations are eliminated with presurgical orthodontics and, preferably, after mandibular growth is complete.

When planning a mandibular setback, aesthetic assessment of the soft tissue of the neck is important. In a patient with a large AP skeletal discrepancy and preexisting heavy neck, correction of the deformity entirely in the mandible would likely worsen the appearance of the neck. In this case, either an isolated maxillary advancement or a combination of maxillary advancement and mandibular setback may be considered. In addition, advancement genioplasty and submental or submandibular liposuction or lipectomy should be considered to improve the patient's overall appearance.

Patients who present with mandibular retrognathia usually present with a class II occlusion, which can also have a significant amount of dental compensation. This condition can be seen in isolation, but is sometimes also seen in patients with VME when the mandible rotates clockwise along the axis created by the mandibular condyle.

Treatment of isolated mandibular deficiency usually involves mandibular advancement. Bilateral sagittal split osteotomies (BSSO) with rigid fixation is the most frequently performed procedure. In general, stability of the mandibular advancement is better with smaller amounts of advancements than with large ones (22). Also, concerns of the soft tissue of the neck are less critical in these patients compared with the mandibular setback patients. When advancing the mandible, the soft tissues of the neck and the cervicomental angle tend to show improvement.

The BSSO is a very versatile operation in that it can be used both for mandibular advancement and for mandibular pushback. It lends itself to rigid fixation, which, in turn, means that the patient will not require MMF. The BSSO is an excellent operation for mandibular advancement to about 12 mm. If the advancement required is greater, other options should be considered, because there will be minimal to no overlap between the proximal and distal segment, creating a much greater potential for relapse. Similarly, the operation is very well suited for mandibular setback of small to moderate distance. Beyond 7 to 8 mm, posterior repositioning of the mandible with the BSSO is difficult, and consideration should be given to an inverted "L" osteotomy or IVRO. When it is necessary to position the mandible a great distance posteriorly, the proximal aspect of the distal segment will cause interference and it might be necessary to remove some of the bone with either a bur or a rongeur. Asymmetry cases must be carefully evaluated. Minor asymmetries can easily be managed with a BSSO (Fig. 8).

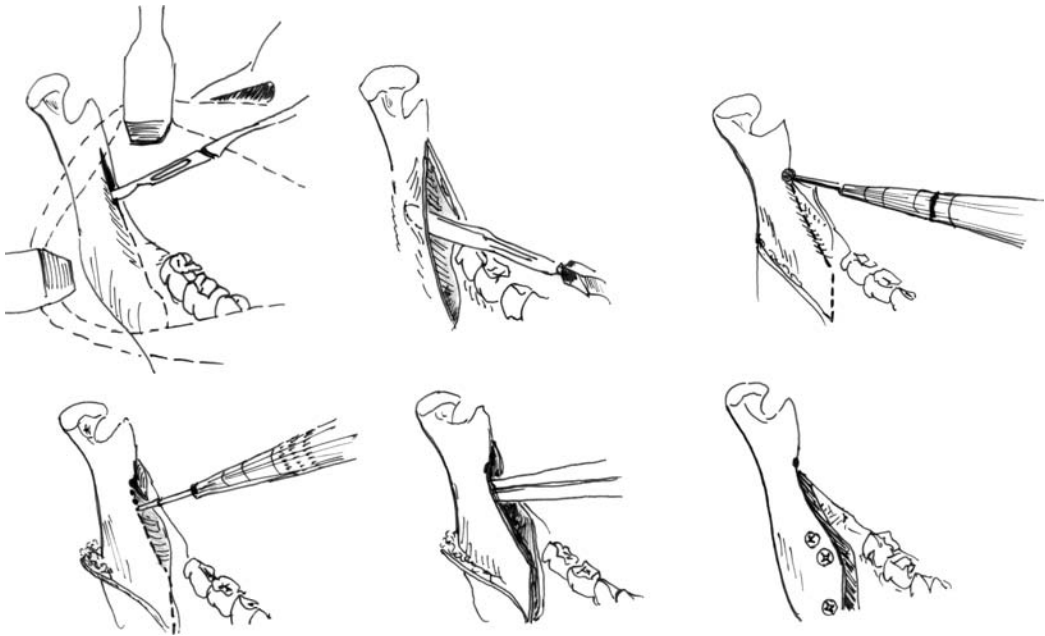


FIGURE 8 Steps in a mandibular bilateral sagittal split osteotomy.

BSSO Technique

The patient is placed in a supine position and nasotracheal intubation is performed. The tube is secured to the nasal septum as previously described for the maxillary osteotomy. Bilateral inferior alveolar and long buccal nerve blocks are given utilizing a local anesthetic with a vasoconstrictor. Local infiltration should be performed on either side of the mandibular ramus as well as in the buccal vestibule along the molar and premolar region of the mandible.

Electrocautery or a knife is utilized to make an incision extending along the anterior border of the ramus and continuing forward along the external oblique ridge. This incision is carried forward up to the level of the first molar. The incision is made through periosteum to the level of the bone.

A periosteal elevator is used to dissect all attached tissues along the entire buccal surface of the ramus and proximal body of the mandible. The dissection is carried all the way to the inferior border of the mandibular body and the posterior border of the ramus. A curved retractor is engaged in the inferior body of the mandible. All attachments are removed along the inferior border of the mandible and posterior border of the ramus by running this retractor the full length of the incision. A V-shaped retractor is then placed on the external oblique ridge and all attachments to the anterior border of the ramus are dissected free, as high up along the coronoid process as possible. At this point, it is wise to utilize a curved Kocher clamp at the tip of the coronoid process to keep the soft tissues out of the way.

The next area to dissect is along the medial aspect of the distal ramus. It is very important to make the dissection strictly subperiosteal as to elevate the inferior alveolar nerve with the soft tissue flap. Starting superiorly with the rounded edge of a periosteal elevator, the instrument is passed posteriorly and inferiorly until just superior and posterior to the lingula.

Once all the soft tissue dissection is completed, the osteotomy can begin. A small periosteal elevator is inserted along the medial aspect of the ramus and it is utilized to keep the soft tissue pedicle away from the area of the osteotomy. A reciprocating saw is then inserted between the periosteal elevator and the medial aspect of the ramus. The saw orientation should be parallel to the occlusal plane and just superior to the lingula, which is usually located approximately 1 cm superior to the occlusal plane and one-half to two-thirds the distance from

anterior to posterior of the mandibular ramus (24,25). Understanding the anatomy of the inferior alveolar nerve as it enters the mandible is important in order to avoid injury to this nerve during this step of the osteotomy.

The medial cut is made through the cortical bone just into the cancellous bone along the entire aspect of the ramus between the lingual and the sigmoid notch. The cut is then extended anteriorly along the external oblique ridge to the area of the second molar. This cut can also be made with the reciprocating saw, although some surgeons prefer a fissure bur. Lastly, the reciprocating saw is utilized again to make the final cut, extending from the area of the second molar directly inferiorly to the inferior border of the mandible. This cut should also be through cortical bone only.

Once all the cuts are completed and connected, thin curved osteotomes are utilized to finish the osteotomy. The osteotomes are positioned anteriorly at first, oriented in such a way as to hug the cortical bone. With a Channel retractor in place, the osteotome should be utilized to complete the cut until the metal to metal sound is noted, as the osteotome completes the cut in the inferior mandibular rim. The osteotomes should be progressively positioned posteriorly until the osteotomy is complete. Once this is accomplished, a periosteal elevator is utilized to separate the osteotomy segments. If the segments do not separate easily, it is wise to check and ensure that all the cuts are complete. It is not recommended to force the segments apart with too much force, as this could lead to adverse fractures. While the segments are separating, it is also important to check the position of the inferior alveolar nerve and to ensure that it is attached to the distal segment. If the nerve is hung up on the proximal segment, gentle manipulation with a smooth periosteal elevator will usually allow its release. If the nerve is encased in cortical bone, sometimes small osteotomies are necessary to release it. Once the segments separate, it is important to check that they are completely free of attachments to one another and that the condyle is indeed attached to the proximal segment.

At this point, the splint is utilized to position the mandible in the desired final occlusion. If the mandible is being positioned posteriorly, it is often necessary to remove some bone from the distal aspect of the proximal segment to allow for adequate bone apposition of the two segments. If the mandible is being positioned anteriorly, there will be a gap between the segments and no bone removal will be necessary.

Fixation of the two segments is done by utilizing three bicortical screws on either side. When this is done, the condyles must be properly seated in the glenoid fossa. It is usually helpful to have the assistant hold the proximal segment in its proper position with a Kocher or hemostat while the fixation is being performed. It is also important to align the inferior border of the mandible of the two segments, as this will eliminate torque of the segments around the osteotomy site. Although there are multiple configurations as to where to place the fixation screws, perhaps the easier placement is two screws superiorly and one inferiorly. It is important in placing the screws to ensure that they are bicortical and that they do not injure the inferior alveolar nerve. Although most times, the screws are placed via a transcutaneous approach utilizing a trochar technique, it is also possible to place the screws via an intraoral technique utilizing special retractors and a right angle drill and screwdriver. Most surgeons routinely use rigid fixation to avoid MMF. Other fixation options such as MMF and even skeletal suspension wires should still be discussed with the patient, especially when large advancements are performed and there is little apposition between the segments (Fig. 9) (26).

Chin Deformities

The chin is a small element of the facial structure. However, it occupies a prominent position, which has made it an important aspect of facial balance and an object of admiration for centuries. Facial contouring surgery is an important part of a plastic surgery practice. Its goal is the correction of the bony tissues of the face to obtain facial harmony (27,28). This harmony is mainly determined by the size, shape, position, and proportion of the chin in respect to the other facial elements (29,30).

The contour of the chin is determined by the contributions of its osseous and soft tissues. The excess or the deficiency of one or more elements could create a facial imbalance.

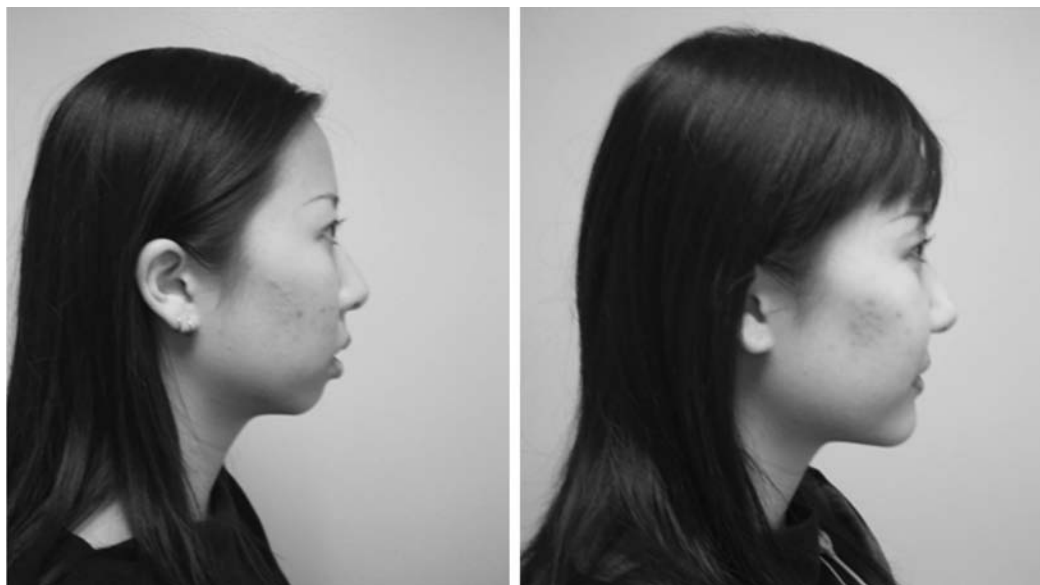


FIGURE 9 Patient with class II occlusion after bilateral sagittal split osteotomy mandibular advancement.

Furthermore, the position and form of the chin determines the position and function of the lips, since many of the muscles of perioral facial mimics have their origin or insertion in the chin. Techniques that modify the position of the chin provide relaxation of the perioral musculature and improve the profile of the lower lip and the labial competence.

Patients with complaints about their chin should be evaluated systematically, as any other orthognathic patient. It is important to confirm that, from the anterior view, the chin position is midline. It is also important to take into consideration the facial proportions as a whole. The facial thirds can give the surgeon clues as to whether the vertical dimension of the chin will need to be addressed. Checking the patient's occlusion is also extremely important as a percentage of patients who complain of a small chin actually suffer from retrognathia and will need mandibular advancement. In patients with adequate facial proportions and a class I occlusion, attention should finally be directed to the chin itself.

The techniques that can be used to change chin position and/or size fall into two main categories. One involves bony osteotomies while the other uses different alloplastic implants. This chapter will only discuss the osteotomy option.

The indications for genioplasty include positioning of the chin in the facial midline and alterations in the vertical or horizontal position of the chin in order to achieve better facial balance. A geniopalasty can be done as a single procedure or in conjunction with orthognathic surgery as part of an overall treatment plan.

Genioplasty Technique

Although this technique can be performed with local anesthesia and sedation, it is probably best done with general anesthesia. The patient should be positioned supine. A throat pack is advisable. The surgeon should be positioned at the head of the table, as this will give the best view of the surgical field and the best perspective as to the location of the midline.

Local anesthesia with a vasoconstrictor should be infiltrated along the entire labial vestibule of the anterior mandible, extending posterior until the area of the second premolar. A surgical knife or Bovy should be utilized to make an incision of the free mucosa of the vestibule, extending from the area of the cuspids on one side to the cuspids on the contralateral side. Although some surgeons prefer to make the incision into the lip and dissect through the mentalis muscle fibers to reach the bone, others prefer to make the incision close to the alveolar

bone, leaving a minimum amount of free mucosa to suture to, which allows for less deinervation of mentalis muscle fibers.

The incision should be carried deep to bone and a periosteal elevator should be utilized to dissect all soft tissue free from the chin along the anterior aspect of the mandible extending to the inferior border of the anterior mandible. It is important to dissect along the anterior aspect of the inferior border of the mandible until and beyond the location of the mental foramen. The inferior alveolar nerve exits the body of the mandible through the mental foramen located in the region of the second premolar approximately bisecting the vertical dimension of the mandible at this point. Once the neurovascular bundle is identified, it is important to dissect inferior to it, as the genioplasty osteotomy extends posterior on the mandible to this point.

Once adequate bony exposure is achieved, it is important to mark the midline. Although this can be done with a pencil or marking pen, most surgeons prefer to use an oscillating saw to make a thin indentation on the bone. The oscillating saw is then utilized perpendicular to this line and at the level of the B-point, well beyond the apices of the incisal roots, to make an osteotomy of the anterior mandible's full thickness through the lingual cortex. The reciprocating saw is then utilized to complete the osteotomies laterally. When extending the osteotomies laterally, it is important to keep the cut at least 5 mm inferior to the mental foramen to prevent injury to the neurovascular bundle. When cutting in the area of the inferior border to the mandible, the surgeon should palpate the saw blade tip along the inferior border by placing a finger of the nondominant hand on the outside of the face in the area of the osteotomy. It is important to ensure that both cortices have been cut. When all the cuts are complete, a periosteal elevator is placed at the osteotomy site in the midline and a torquing motion is applied in order to separate the two segments of bone.

Once the two segments separate, the distal segment will still have its lingual attachments consisting of the genioglossus and geniohyoid muscle. It is important to stretch these muscles if an advancement is anticipated. The distal segment is then placed in the desired position and fixation is applied. Before fixation is performed, however, it is helpful to smooth out any contour irregularities or sharp bony edges at the osteotomy site, especially on the proximal segment. Fixation of the segments can be done with prefabricated plates of different lengths or with bicortical 2 mm screws. Usually one or two screws are sufficient for proper fixation.

When vertical dimension changes are anticipated, it is important to design the osteotomies for maximum efficiency and to optimize the technical feasibility of the procedure. If a reduction of the vertical dimension is anticipated, a superior osteotomy is marked at the area of the B-point in the mandible, and a parallel osteotomy is marked inferior to the original one. The distance between the two parallel osteotomies should be equal to the planned reduction of the vertical dimension. It is important to perform the inferior osteotomy first, and then the superior one, as it would become extremely difficult to perform the inferior osteotomy after the distal segment has been cut and has become very unstable. Once both osteotomies are complete, the intervening bone is removed and the two segments are placed in apposition. Fixation is then performed. If the vertical dimension is to be increased, only a single osteotomy is necessary and an intervening bone graft is placed between the two bony segments.

Once the fixation is completed and after copious irrigation, the soft tissue is closed. Although this can be done in two layers (muscle and mucosa), a single layer with a resorbable suture such as 4-0 Chromic should suffice. External dressing is not necessary, but some surgeons prefer the use of foam tape around the chin (Fig. 10).

Depending on the variations in techniques, the goal of the procedure, and where and how the distal segment is positioned, the basic genioplasty technique is given different names (31):

1. *Sliding genioplasty*: The osteotomy segment slides anteriorly, and the lower facial third height is modified minimally or not at all.
2. *Jumping genioplasty*: The caudal segment is placed over the anterior portion, in front of the mandible, almost as an implant. However, the lower soft tissue attachments of the segment should be preserved to avoid resorption. This name was used by Tessier and it is indicated to improve the sagittal projection of the chin and decrease the height of the lower facial third.



FIGURE 10 Steps in performing a genioplasty.

3. *Graft genioplasty or interpositional genioplasty:* It is used when the amount of material is inadequate and it is necessary to advance the chin as well as increase the lower facial height. In these cases, the advancement is achieved with the segment and the facial height with a bone graft or a hydroxyapatite block in between the segments.
4. *Wedge genioplasty:* This type of genioplasty helps increase the AP projection of the chin and decrease the height of the lower facial third. Two horizontal cuts parallel to each other and the occlusal plane are done. The caudal cut is made, then the segment between the cuts is resected. It corresponds to the amount of height to be reduced in the lower facial third. Patients operated with this technique should know that some amount of soft tissue ptosis will occur, resulting in crowding of the submental space.
5. *Oblique genioplasty:* This technique advances the chin in the sagittal plane and reduces its height without taking out a bone fragment. The steeper the cut, the bigger is the reduction in chin height.
6. *Stepladder genioplasty:* This technique is used in cases of an important sagittal advancement, without modifying the lower facial third height. Two parallel cuts are done and the lower segment is advanced over the higher segment, so a stepladder is created.
7. *Asymmetric genioplasty:* These are a special group of procedures indicated in patients with vertical and/or horizontal asymmetries of the chin. In this surgery, a lateral wedge of bone is resected from the longer side and used in the contralateral shorter side. Also, the midline is shifted and centered with the facial axis during the procedure.

Combined Deformities

Some patients will present with combined maxillary and mandibular abnormalities. The typical case is the patient with VME and mandibular retrusion or prognathism. The correct diagnosis of these patients is extremely important, and cephalometric analysis and dental casts are extremely helpful. Once the preoperative orthodontics is completed, it is important to repeat a cephalogram and a set of dental casts from which to formulate the surgical treatment plan. On this type of patient, the models will need to be mounted via a facebow transfer onto a semiadjustable articulator prior to performing the model surgery. The maxillary movements are determined primarily from clinical evaluation and the cephalometric analysis. After the intermediate splint is fabricated, the mandible is positioned in the correct occlusion and the final splint is made. The chin position must then be assessed during the operation and after the mandibular osteotomy is performed and the mandible fixated. In a great number of these double-jaw cases, a genioplasty must also be done in order to place the chin in proper facial balance. The mandibular movement is used to achieve optimal occlusion, and, if necessary, a genioplasty is added in order to achieve proper chin position and facial harmony (Fig. 11).

CONCLUSION

A certain number of patients with facial cosmetic complaints have skeletal and occlusal abnormalities that are the source of, or greatly contribute to, their chief complaint. Thus, it is



FIGURE 11 Patient with vertical maxillary excess and mandibular prognathism before (*left*) and after (*right*) Le Fort I maxillary impaction and bilateral sagittal split osteotomies mandibular pushback.

important for the plastic surgeon evaluating these patients to be familiar with the tools used to evaluate the orthognathic surgery patient. The correct diagnosis of the etiology of the patient's problem is of paramount importance for adequate treatment and to achieve optimal results. Even if the patient is not interested in orthognathic surgery and is seeking "camouflage" surgery, the surgeon should discuss all of the treatment options and explain the pros and cons of each of the treatment modalities.

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14 Craniofacial Distraction Osteogenesis

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INTRODUCTION

Distraction osteogenesis has become an important technique for craniofacial surgery over the last 18 years. Recent research and developments have centered around (i) identifying appropriate indications for the distraction procedures, (ii) perfecting distraction instrumentation and devices, and (iii) understanding the biology of distraction within membranous bone (1–3). For the craniofacial surgeon, distraction osteogenesis has been a bridge towards tissue engineering techniques. It offers the benefit of reduced morbidity without a need for bone grafting. In addition, there is the theoretic improvement of decreased relapse from less soft tissue recoil postprocedure due to the gradual lengthening of the distraction technique.

HISTORY

The history of distraction osteogenesis is seated in the history of fracture repair with the use of continuous traction for the reduction of displaced fractures (4). Distraction osteogenesis techniques have been modified from the traditional techniques of osteotomies and bone fixation (5). Early in the 20th century distraction lengthening in the lower extremity was fraught with problems from infectious complications and bony nonunion. Beginning in the 1950s, Ilizarov made great strides in optimizing the distraction process in lower extremities with his external ringed appliances in over 15,000 cases (6).

History of distraction in the craniofacial skeleton began with animal experimentation on the mandible in the 1970s; however, it was not until 1989 that McCarthy first began the clinical use of distraction with human mandibular lengthening (7,8). Since then, distraction has been successfully performed on patients with various craniofacial hypoplasias (craniofacial microsomia, Pierre–Robin sequence, Treacher–Collins syndrome, Nager syndrome), temporomandibular joint ankylosis, craniofacial dysostosis (for midface and forehead deficiencies), posttraumatic deformities, and on most bony regions of the craniofacial skeleton (mandible, maxilla, zygomatic/malar, orbital, frontal/forehead, parietal and occipital, sphenoidal, and even the cranial base) (1).

BIOLOGY

Similar to fracture healing, distraction osteogenesis involves: (i) an initial injury (the osteotomy), (ii) a recruitment of cells (including mesenchymal stem cells and preosteoblasts), (iii) a mechanical linear force (from the distraction device to induce and direct the formation of both hard and soft tissues), and (iv) callus formation (mineralization of bone with consolidation). The term osteogenesis (formation of vascularized bone *de novo*) should be differentiated from the terms osteoconduction (creeping substitution of new bone into the peripheral regions of an implant) and osteoinduction (new bone induction with the use of growth factors) (9).

There are three phases of distraction: *latency*, *activation*, and *consolidation* (10). Latency is the period immediately following the osteotomy. Typically, the latency for craniofacial distraction is anywhere from 0 to 2 days, except with a monobloc distraction procedure where a one-week latency period is beneficial (to allow for mucosal healing between the subdural space, “clean,” and nasal sinuses, “colonized,” before beginning distraction).

Activation involves *rate, rhythm, length, and molding*. A standard rate of distraction is 1 mm per day. Any major variation of this may result in fibrous union (rate is too fast) or premature consolidation (rate is too slow). (However, with neonatal distraction faster rates of up to 2 mm per day are necessary) (11). A rhythm or frequency of turning the distractor arm two times per day is often used; however, it is yet to be determined scientifically whether this rhythm is superior to a more frequent interval or even continuous distraction. The length of distraction may often be determined preoperatively and is predicted by the size of the defect or amount of length necessary to correct a functional problem (e.g., upper airway obstruction). Molding the new “generate bone” is possible because of the plasticity of the region prior to consolidation. Molding may be done to close an open bite, correct form, or improve symmetry. Guiding the generate bone may be done with multi-vector distractors, dental elastic bands, or other external force.

Consolidation involves the mineralization and hardening of the new bone formed by the distraction osteogenesis process. The study of zones in the distraction site show that the peripheral zones (close to the osteotomy sites) begin and end consolidation sooner than the central zones (10). The time of full consolidation may vary but is generally between 6 and 12 weeks following the completion of distraction. Internal distraction devices are often kept in place for three months. External devices are usually removed at six weeks because of their cumbersome nature. Slower mineralization and the need for longer consolidation time occurs in older patients, patients with prior exposure to radiation, and patients who develop infections (12).

MANDIBULAR DISTRACTION

Mandibular distraction allows for (i) the formation of vascularized mandibular bone to correct hypoplasias and (ii) the expansion of the regional soft-tissue envelope. Mandibular distraction is commonly used in growing patients with skeletal deformities; however, may also be used in skeletally mature patients who require extremely large advancements. The use of conventional techniques with large advancements (> 10 mm) may result in considerable relapse and undue stress on the inferior alveolar nerve (13). In general, for skeletally mature patients, conventional techniques for mandibular advancement (like orthognathic: bilateral sagittal split osteotomy of the mandible, etc.) are preferred. The advantage to an orthognathic procedure is that a patient may obtain optimal occlusion in just one operation.

Indications for mandibular distraction include: micrognathia (with upper airway obstruction, obstructive sleep apnea, or tracheostomy dependency), mandibular asymmetry (craniofacial microsomia) and severe class II malocclusion (Table 1). The modified Pruzansky classification system for bilateral or unilateral mandibular hypoplasias is useful in describing severity seen radiographically and in planning treatment options (14,16). Grade 1 mandibles have normal configuration but have a reduction in ramal and condylar size. Grade 2a mandibles have a small, malformed condyle but the relation to the glenoid fossa is maintained. Grade 2b mandibles have an abnormal temporomandibular joint. Grade 3 mandibles lack a condyle, ramus, and glenoid fossa.

Devices and Vectors

Preoperative preparation for mandibular distractions involves choosing an appropriate device (external or internal) and planning the length and vector of distraction. Preoperative radiographic evaluations may include a panoramic radiograph, lateral cephalogram and a

TABLE 1 Indications for Mandibular Distraction

Micrognathia
Upper airway obstruction
Obstructive sleep apnea
Tracheostomy dependency
Mandibular asymmetry
Craniofacial microsomia
Sever class II malocclusion (skeletally mature)

three-dimensional computed tomography scan (particularly for visualization of the temporomandibular joint region).

External mandibular devices were the first designed and used devices. They offer the advantage of ease of placement and removal. However, external devices are more likely to become dislodged during the distraction process. As mentioned above, shorter consolidation times have been used because of the cumbersome nature of the devices. Although a multiplanar device offers the ability to adjust the distraction vector, less precision may occur because of the increased distance from the body of the device to the osteotomy and because of pin loosening or bending. In addition, with the external devices pin care is necessary and external scarring results from pin movement.

Internal mandibular distraction devices offer the advantage of being hidden from sight so the patients more often return to school during the consolidation phase. Disadvantages include: the application and removal may be technically more difficult, there are more limitations with length and more subperiosteal stripping may be required. Although theoretically this more extensive undermining may decrease blood supply, it has not been shown clinically that bone healing with internal distraction is inferior to external distraction. Internal mandibular devices may be uniplanar, telescopic (a shorter initial rod that lengthens more), have a right angle activation arm (for vertical vector placement), be curvilinear or another design.

The distraction vector may be horizontal, vertical, or oblique. The position of the distraction device not the osteotomy position determines the vector. A horizontal vector is chosen for bilateral mandibular body deficiencies, like Pierre–Robin sequence. A vertical or an oblique vector is important for lengthening the mandibular ramus in most other cases, as in Treacher–Collins syndrome or craniofacial microsomia. After mandibular lengthening, before consolidation, the generate bone may be molded with external forces (elastic bands) to optimize the final distraction vector.

Mandibular Distraction Operative Technique

1. An oral-ray endotracheal tube may be used and is suture-secured interdentally.
2. After 0.25% marcaine and epinephrine injection, a mandibular gingivobuccal sulcus incision is performed with a Colorado-tip bovie.
3. Subperiosteal elevator, “J-stripper” (to remove masseteric mandibular angle attachments), “Y-stripper” (to dissect the coronoid process) are used for subperiosteal dissection of the mandibular ramus and body.
4. A channel retractor is placed for exposure and a Kocker with a chain is clamped onto the coronoid process. The subperiosteal dissection is completed with a curved elevator on the lingual aspect of the ramus above the lingua (proximal to entrance of inferior alveolar nerve).
5. The mandibular osteotomy is marked with a Colorado-tip bovie from the anterior aspect of the ramus just above the lingua, obliquely down the buccal aspect of the ramus to the inferior mandibular border just anterior to the angle. (Preservation of enough bone proximally is important for easy fixation of the distractor plates. As with all osteotomy procedures of the mandible, extreme care is taken to avoid injury to the inferior alveolar nerve.)
6. Reciprocating saw, directed parallel to the occlusal plane, is used for a corticotomy of the lingual aspect of the ramus above the lingua. Osteotomy is continued through the anterior aspect of the ramus but in a bicortical fashion. Next, a bicortical osteotomy is performed through the thick inferior border of the mandible and a corticotomy is extended up the buccal aspect of the ramus to connect with the previous osteotomy. Greenstick fracture is performed with a periosteal elevator, or curved osteotomies may be used to complete the osteotomy as in a sagittal split osteotomy.
7. Distraction devices are fashioned. (For internal devices, plates are customized and appropriate length turning arm is applied.) Intraoral or percutaneous placement of 2-0 fixation screws are used. (High profile screws are recommended for ease of removal following consolidation.) Turning arms are brought out through the intraoral incision to avoid external scarring; however, percutaneous exit of the turning arms is also an option.
8. Distraction devices are tested in situ and repositioned to a “zero” of undistracted state.
9. Oral mucosa is closed with a running locking 4-0 chromic suture.

Postoperative care for mandibular distraction patients is similar to other orthognathic procedures with a soft diet. As mentioned above the distraction activation or turning is begun the next day (minimum to no latency), the frequency is twice daily at a rate of 1 mm per day until completion of distraction length. Intraoral distraction turning arms are then removed and anterior elastic bands are placed on brackets, or on maxillomandibular fixation (MMF) screws to mold the generated bone and close the open bite. At times a Panorex or lateral cephalogram is taken during or at completion of distraction to monitor the process. Both devices are removed after three months of consolidation.

NEONATAL DISTRACTION

The purpose of neonatal mandibular distraction is to correct upper airway obstruction to avoid a tracheostomy. Although a tracheostomy may be life saving for a newborn with micrognathia, a tracheostomy may also be complicated by tracheitis, pneumonia, laryngomalacia, bleeding from stomal granulation, subglottic stenosis, or long-term problems, such as, developmental and speech delays. Distraction lengthening of the mandible corrects posterior tongue collapse and elevates the epiglottis, alleviating the need for even a temporary tracheostomy. Once a tracheostomy is placed in a newborn it may take years and multiple surgical procedures (such as laser ablation for tracheomalacia and tracheal reconstruction) to remove it.

Selection of appropriate candidates for neonatal distraction involves a multidisciplinary approach and preoperative tests. The multidisciplinary team always includes a neonatologist, a plastic surgeon, an anesthesiologist, and head and neck surgeon; and often includes a pediatric pulmonologist, pediatric gastroenterologist, and geneticist. Patients considered candidates for neonatal distraction have severe micrognathia and upper airway obstruction (Fig. 1). Patients excluded as potential candidates for neonatal distraction require a tracheostomy and have (i) other airway lesions, like a tracheal web, (ii) central sleep apnea, or (iii) severe gastroesophageal reflux. In addition, neonatal distraction is not required if the obstruction is moderate or mild and may be controlled with prone or side positioning. Tongue-lip adhesion or a secured nasopharyngeal tube may also be used in these cases to control the tongue prolapse temporarily. For mild or moderate airway obstruction cases, these techniques, as well as proper positioning, may allow time for the mandible to have “catch-up” growth. Pierre–Robin sequence (micrognathia, glossoptosis, and cleft palate) is the most common diagnoses of patients undergoing neonatal distraction, but patients with Treacher–Collins, Nager, and other syndromes have also undergone the procedure to avoid a tracheostomy.

Neonatal Distraction Operative Technique

Although an external device may be used, our team prefers an internal device (particularly our neonatologists for postoperative care) (Fig. 2).

1. Open gingivobuccal sulcus and elevate periosteum exposing the lateral mandible.
2. Make a Risdan incision well below the mandibular body approximately 2 cm long.
3. Spread to the periosteum and retract.
4. In the subperiosteal plane connect to the existing pocket.
5. Elevate on the lingual aspect of the ramus and place a malleable for protection.
6. Place a single hook on the angular notch and mark the inverted “L” osteotomy with the bovie cautery.
7. Use a reciprocating saw to complete the osteotomy.
8. Select the internal microdistractor with locking mechanism (to avoid turn-back; KLS-Martin) and fashion the device by cutting off accessory plates and attaching the turning arm.
9. Place the device with the turning arm through the gingivobuccal sulcus incision and plan the orientation (vector). (For Pierre–Robin sequence with a short mandibular body a horizontal positioning is usually performed.)
10. Fixate the device with 1.0 mm × 5.0 mm screws.
11. Distractor function is tested and then returned to the zero or undistracted position.
12. Incisions are closed.

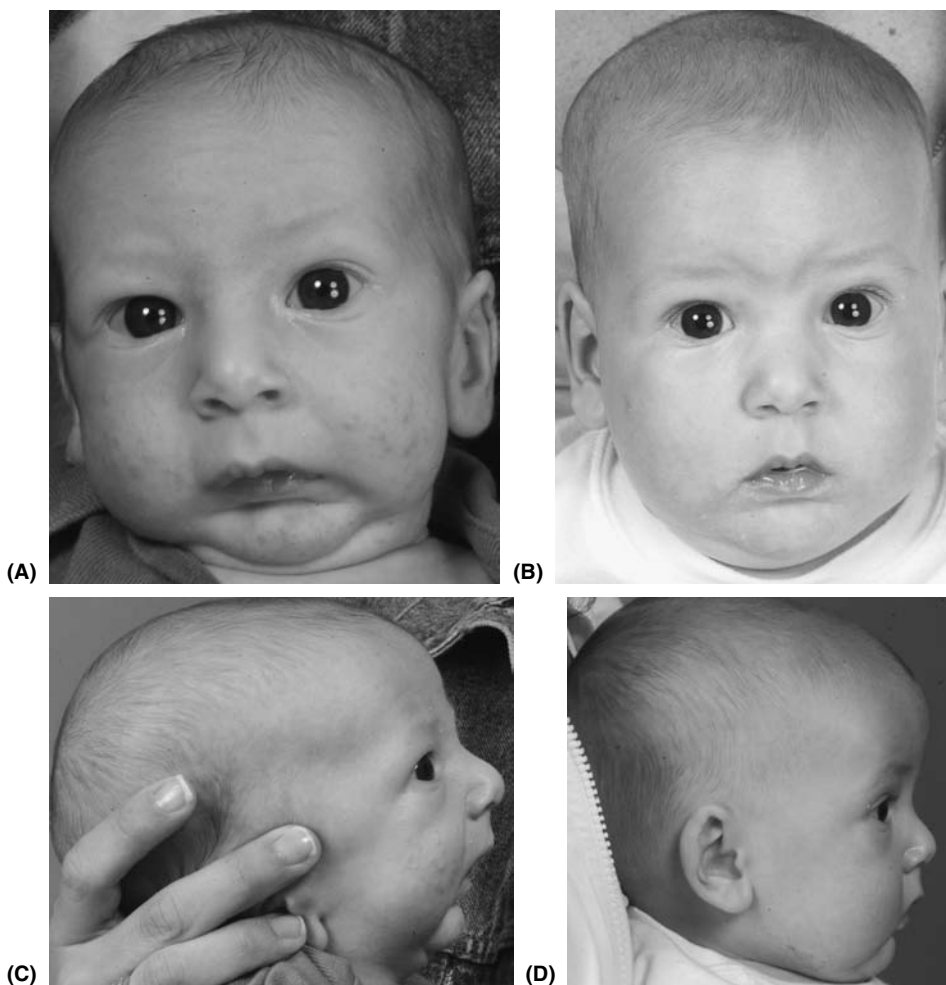


FIGURE 1 Neonatal distraction. (A) Pre-operative image of a patient with Pierre–Robin sequence. (B) Postoperative appearance with bone consolidation. (C) Lateral view of patient with Pierre–Robin sequence prior to distraction osteogenesis. (D) Lateral patient image postoperative. Note the improved chin point and secondary weight gain over three months.

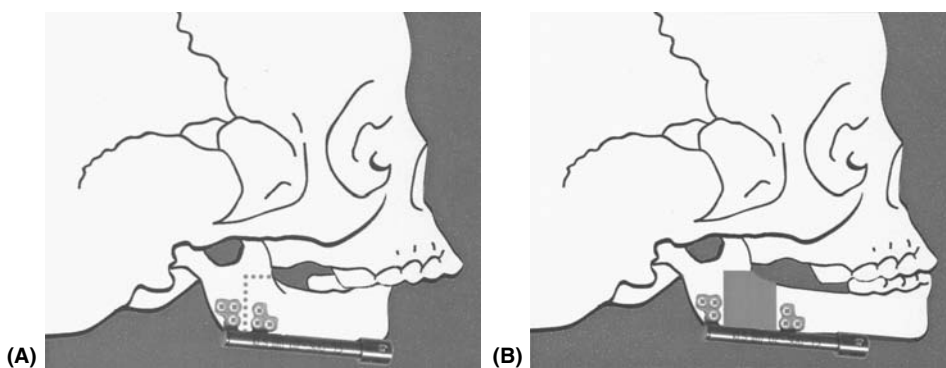


FIGURE 2 Neonatal distraction device. (A) Illustration of inverted L-osteotomy with the initial placement of neonatal distraction device. (B) Illustration of lateral mandible post-distraction. Note the area of consolidation indicated in light grey, and the position of the anterior mandible in relation to the maxillae.

The newborn is transferred to the neonatal intensive care unit or pediatric intensive care unit, intubated and sedated. A frequency of twice daily and a rate of 2 mm per day are used for distraction. After approximately 7 to 10 days the infant returns to the Operating Room for turning arm removal and extubation in a controlled setting. Oral feeding typically progresses over the subsequent 24 days and the infant is discharged. Six to 12 weeks later the intraoral devices are removed through the external Risdan incision as an outpatient.

LE FORT I MAXILLARY DISTRACTION

Le Fort I distraction may be used to treat patients with severe maxillary hypoplasia, like patients with cleft lip and palate with intrinsic maxillary growth disturbances or scarring (Fig. 3) (15). For patients with class III malocclusion and mild to moderate maxillary hypoplasia (<10 mm of advancement required) an orthognathic procedure at skeletal maturity is recommended. However, for patients with class III malocclusion and severe maxillary hypoplasia (>10 mm of advancement required) Le Fort I distraction offers larger advancements with less relapse (16). For patients with alveolar defects, bone graft consolidation must be done prior to a Le Fort I distraction procedure. If the maxillary segment is in more than one piece, the distracted segments cannot be controlled.

Le Fort I Maxillary Distraction Operative Technique

For skeletally mature patients that may require an orthognathic procedure after distraction, at the time of device removal, preoperatively model surgery is preformed and occlusal splints are made.

1. Nasal Ray endotracheal tube is secured and throat pack is placed. Gingivobuccal sulcus incision is made with the Colorado tipped bovie.
2. Subperiosteal dissection is performed with an elevator to deglove the anterior maxilla, ptergomaxillary region, piriform aperture and inside the nasal cavities.
3. Osteotomies are performed with the reciprocating saw at the Le Fort I level through the nasomaxillary and zygomatic maxillary buttresses, as well as the septum. (A high Le Fort I osteotomy, superiolaterally to the infraorbital foramen, is performed for patients with hypoplastic malar regions.) A Kawamoto osteotome is then used for the ptergomaxillary osteotomy.
4. Maxillary down-fracture is performed and the serrated Kawamoto osteotome is used within the ptergomaxillary osteotomy site to stretch the soft tissues.
5. Posterior bony interferences of the Le Fort I segment may be removed with a rongeur or Kerrison grasper.
6. Then, the internal Le Fort I distraction device (KLS Martin L.P., Jacksonville, Florida) is secured with screw fixation. (Plate bending, burring of the maxillary bone for rod positioning, or placement of a red rubber catheter over the turning arm may be done at this time.)
7. The device is tested by advancing the distractor then returning it to the zero position. In addition, elastic bands are placed on hooked brackets or on 4-MMF screws (KLS Martin L.P., Jacksonville, Florida). (These elastic bands may be placed either during the original procedure or when the turning arms are removed following distraction in two to four weeks. The bands help guide the Le Fort I segment into appropriate occlusion during distraction and prevent an open bite deformity.)
8. Finally, a nasal cinch suture is used to avoid alar base flaring and the gingivobuccal sulcus incision is closed with running locking 4-0 chromic suture.

Distraction is begun the day after the Le Fort I osteotomy (latency, 1 day) at 1 mm per day (rate) two times per day (rhythm). After completion of distraction lengthening the patient has the turning arms removed so the entire device remains covered during the three-month consolidation period (Fig. 4). Elastic bands, placed on orthodontic hooks or on

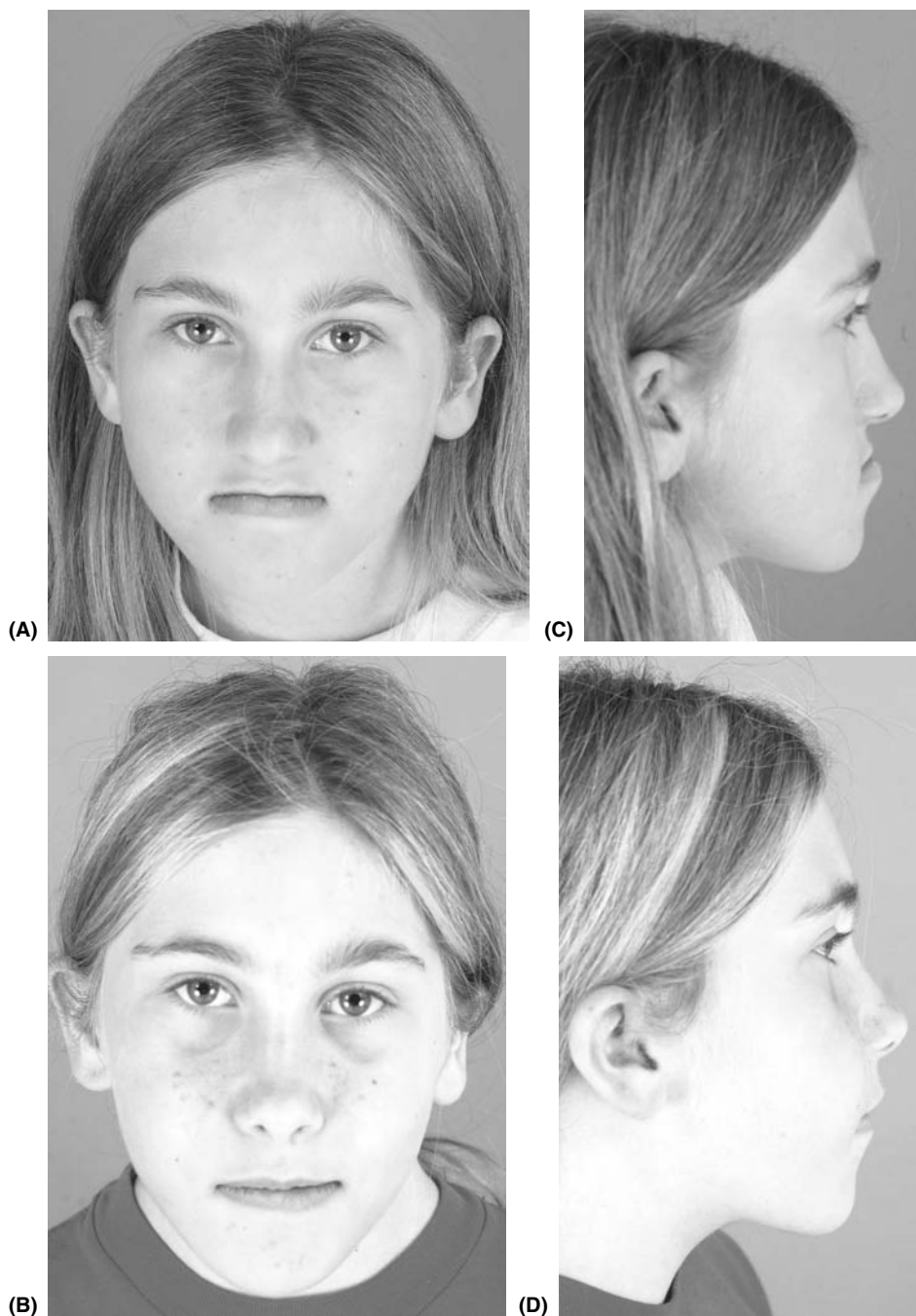


FIGURE 3 Le Fort I distraction. (A) Pre-distraction patient with cleft palate and severe maxillary hypoplasia. (B) Patient postoperative image. (C) Pre-distraction lateral view of patient. (D) Lateral postoperative image after 25 mm Le Fort I internal maxillary distraction.

maxillomandibular fixation screws and hooks, are used to guide the maxillary segment into the desired occlusion. Patients are followed-up weekly until the proper over jet, overbite, and relative stable posterior occlusion are achieved. Distraction devices are removed after a consolidation period of three months.

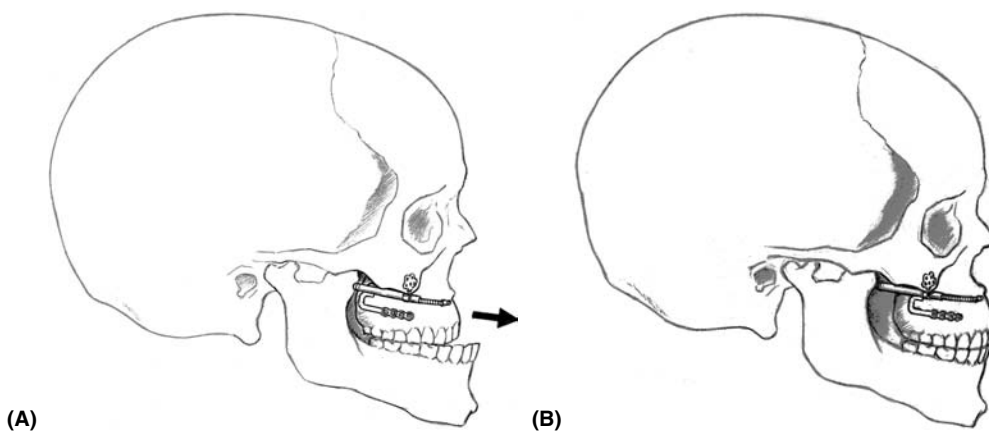


FIGURE 4 Le Fort I distraction device. (A) Illustration of maxillary hypoplasia with internal distraction device placement. The arrow depicts the intended distraction movement. (B) Lateral view illustration of post-distraction with internal device and complete consolidation.

LE FORT III/MONOBLOC DISTRACTION

Correction of patients with syndromic craniosynostosis and midface retrusion may involve either (i) staged procedures with fronto-orbital advancement followed by a Le Fort III advancement, or (ii) a monobloc (one unit) advancement (1–10). Traditionally, these procedures involve bone grafting and rigid fixation. Besides correction of facial deformities, indications for these procedures include upper airway obstruction, obstructive sleep apnea (OSA), tracheostomy dependency and ocular problems (ocular keratitis, corneal ulcers, globe herniation, and blindness) (11–13).

The monobloc procedure is advantageous because it avoids a second major procedure; however, with the traditional one-staged monobloc advancement, the risk of serious complications (meningitis, cerebrospinal fluid leak) is considered too high for many institutions to advocate its use (10,11,13–16). These infections are thought to be caused by the large nasofrontal dead space and communication between epidural and ethmoidal sinus tissues left after the acute advancement of the facial-forehead unit (9,10,14,15). Distraction osteogenesis after a monobloc osteotomy lowers the risk of complications by allowing remucosalization of the nasofrontal area (for 4–7 days) before gradual advancement. Our team prefers the internal monobloc distraction device for both a Le Fort III distraction procedure (Fig. 5) and a monobloc distraction procedure (Fig. 6) because it is more easily tolerated by patients and has consistently good results.

Le Fort III/Monobloc Distraction Operative Technique

1. Zig-zag coronal incision and subperiosteal dissection is used. The temporalis muscle is left down (to avoid temporal hollowing postoperatively) by dissecting just above the superficial layer of the deep temporal fascia (a plane just below the frontal branch of the facial nerve). Osteotomy sites are exposed with subperiosteal dissection around the circumferential orbit, down to the nasofrontal suture and to the zygomatic body.
2. Intraoral exposure is used for the ptergomaxillary osteotomy. Craniotomy of the forehead bone flap is performed and, with retraction of the dura, the orbital roofs are exposed.
3. Osteotomies are performed bilaterally in the anterior zygomatic arch, lateral orbital wall, orbital roof, medial orbital wall, orbital floor, ptergomaxillary buttresses and septum (after the mid-face down-fracture is started). Rowe disimpaction forceps are used to complete the down-fracture. Complete side-to-side mobilization is performed.
4. A pericranial flap (10 cm × 4 cm) may be sutured into the midline defect to separate the nasal sinus cavities from the epidural dead space and Fibrin glue may also be used over the flap.
5. Finally, an internal distraction device (Kawamoto Distractor, KLS Martin) is secured anteriorly to the zygomatic body, just in front of the osteotomy site. Posteriorly, it is fixed

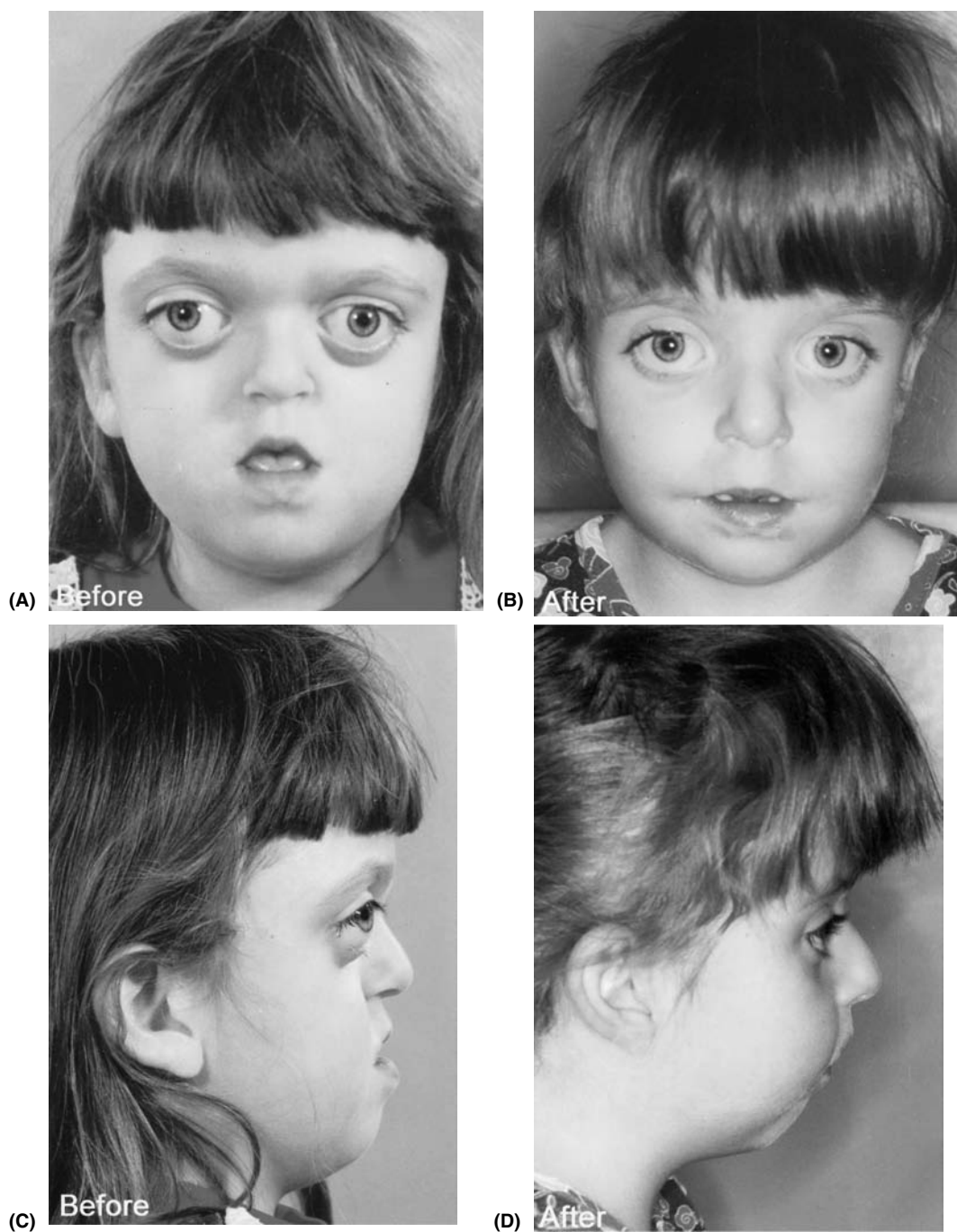


FIGURE 5 Le Fort III distraction. (A) Frontal view of patient with midface hypoplasia prior to distraction. (B) Frontal view of patient after distraction and consolidation. (C) Lateral view of patient prior to distraction. (D) Lateral view of patient after distraction. *Source:* Courtesy of Dr. H. K. Kawamoto.

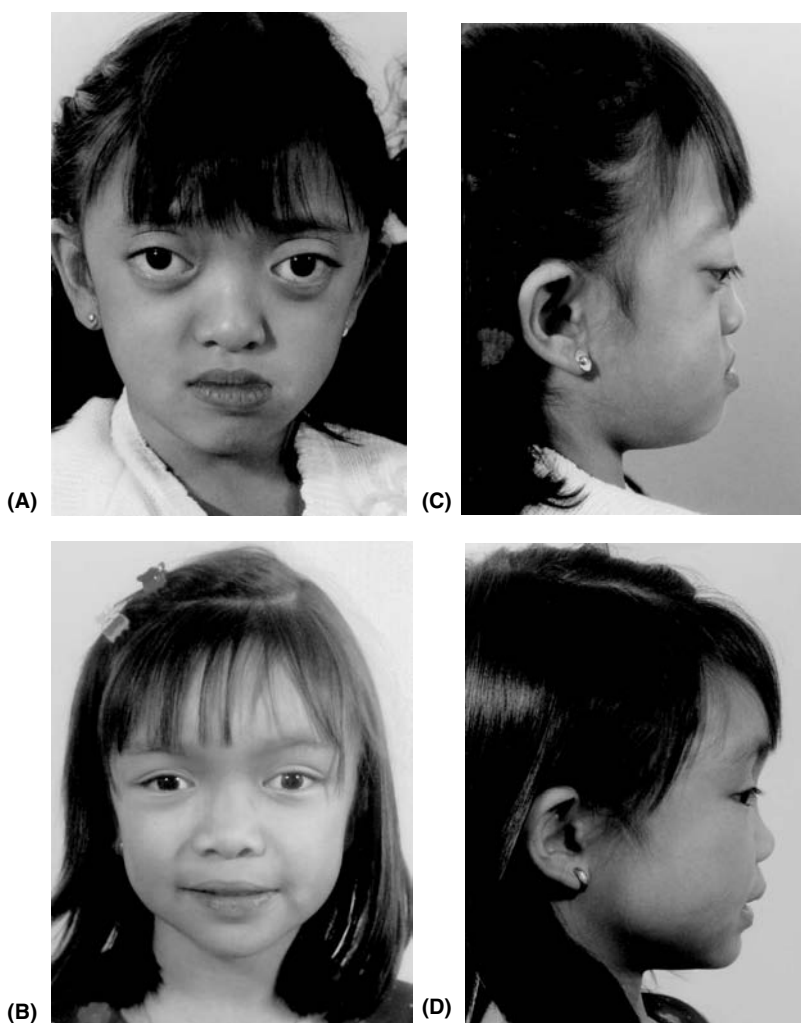


FIGURE 6 Monobloc distraction. (A) Frontal view of patient with forehead and midface deficiency prior to distraction. (B) Frontal view after distraction and consolidation. (C) Lateral view of patient prior to distraction. (D) Lateral view after distraction. *Source:* Courtesy of Dr. H. K. Kawamoto.

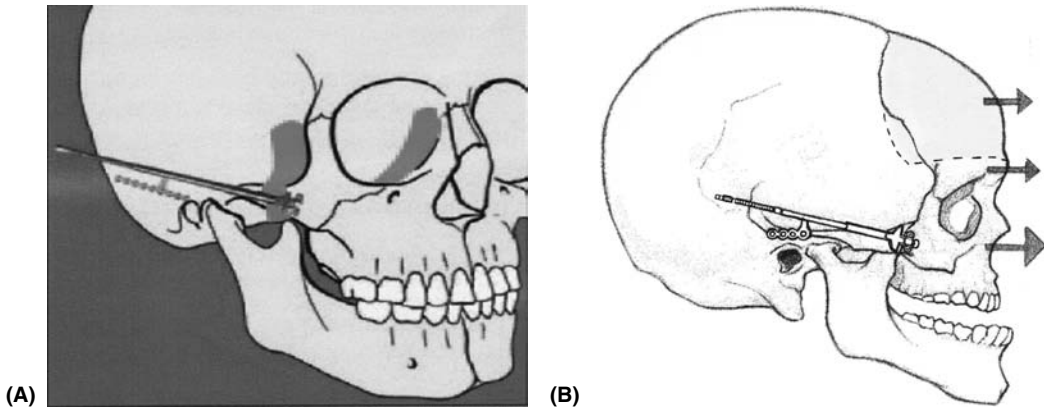


FIGURE 7 Illustration of mid-face distraction. (A) Le Fort III distraction with internal device after advancement, with new bone depicted in light grey. (B) Monobloc distraction, with internal device in pre-distraction position depicting the forward movement of the forehead and the mid-face.

to the temporal bone just behind the zygomatic root and above the external auditory canal (Fig. 7). Distractors are tested and then returned to the start, or zero position. All incisions are then closed over drains.

Monobloc distraction is not started until postoperative day 7 (latency, 7 days). Distraction is performed with an advancement of 0.5 mm twice daily for a total advancement of 1 mm per day (rate, 1 mm/day; rhythm, 2 times/day). For all patients advancement is planned to correct orbital and forehead deformities. Therefore, optimal occlusion is not the primary goal of the procedure. Turning arms are removed after completion of distraction. Elastic rubber bands may be placed on MMF screws to guide the distraction segment and avoid an anterior open bite.

Patients should undergo a consolidation phase of three months. Internal distraction devices are then removed in the operating room through the supraauricular portion of the old incision (4 cm) and a percutaneous cheek incision. Coronal incision and flap turn-down are not necessary for device removal.

For craniofacial dysostosis patients with hypertelorbitism and midface hypoplasia, a simultaneous facial bipartition and monobloc distraction may be performed (Fig. 8).

TEMPOROMANDIBULAR JOINT

Congenital temporomandibular joint (TMJ) bony ankylosis is a rare pediatric condition that poses a great surgical challenge. The surgical treatment goal is to both release the joint ankylosis and lengthen the mandible with the hope of restoring and maintaining normal TMJ movement and mandibular function. Many protocols have been used successfully to treat fibrous TMJ ankylosis from trauma or other acquired causes including, resection gap arthroplasty, and reconstruction with costochondral graft or prosthesis (17). But with bony TMJ ankylosis, this more severe case requires more aggressive treatment with distraction osteogenesis. For these cases of congenital bony TMJ ankylosis, transport distraction osteogenesis has been used with success.

The initial procedure involves mandibular lengthening with distraction osteogenesis to correct micrognathia. Either an internal device (KLS Martin, Jacksonville, Florida) or an external device (KLS Martin or Synthes-Paoli, Pennsylvania) is used depending on the severity of the micrognathia (Figs. 9 and 10). In severe cases, an external device is used (Fig. 11).

After correction of micrognathia with distraction, the transport distraction procedure is performed. This stage involves (i) bilateral resection of the fused condyle, (ii) reshaping of the glenoid fossa, (iii) transfer of temporoparietal fascial (TPF) flap, and (iv) transport of a mandibular ramus segment (Fig. 10B). As the segment of mandibular bone is transported a cartilaginous cap is formed at the leading edge which results in a "neocondyle."

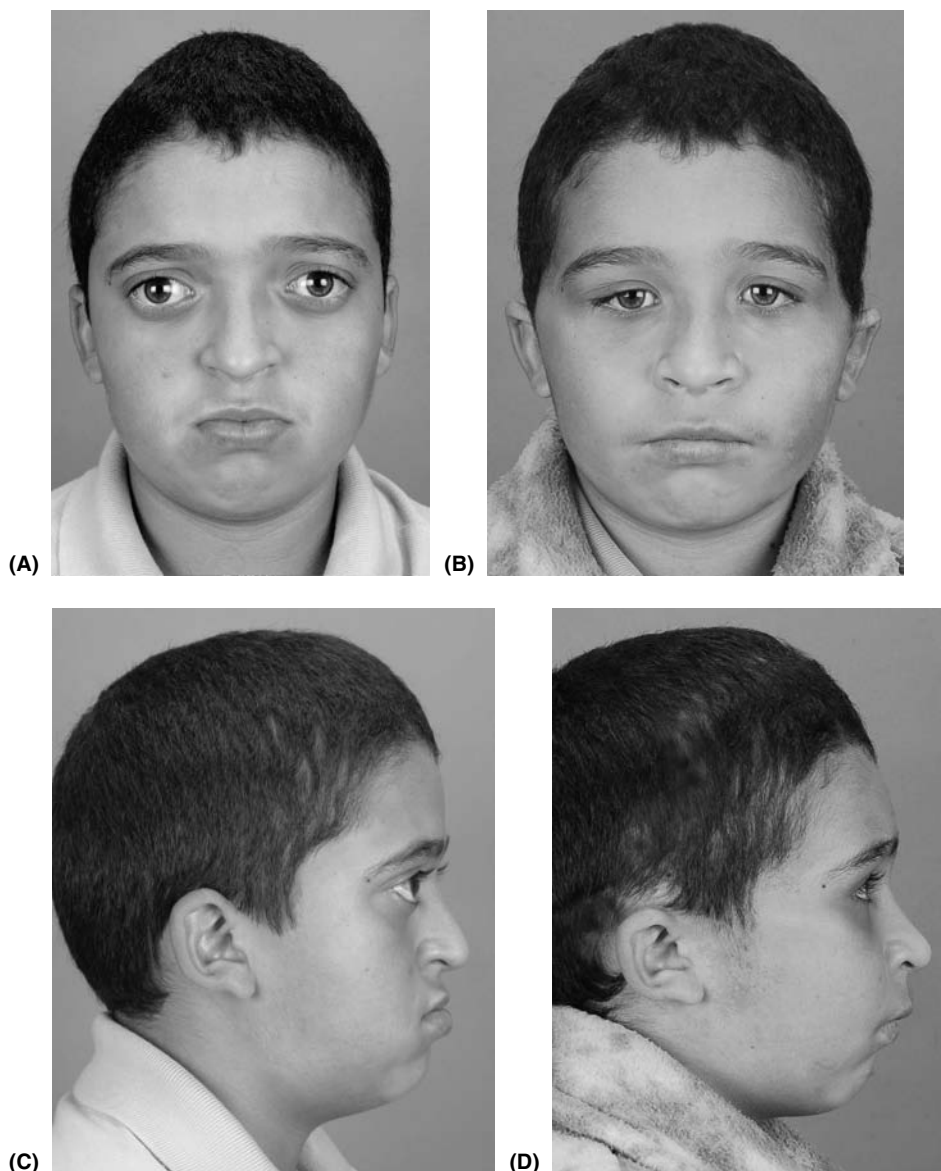


FIGURE 8 Images of Cruzon patient who underwent simultaneous facial bipartition and monobloc distraction to correct both hypertelorbitism and midface hyperplasia. (A) Preoperative frontal view. (B) Postoperative frontal view. (C) Preoperative lateral view. (D) Postoperative lateral view.

Transport Distraction Operative Technique

1. Through a zig-zag temporal hairline and preauricular incision, dissection is performed down to the TPF and zygomatic root. A pedicled 2 cm × 5 cm TPF flap is raised.
2. Using a facial nerve stimulator, bipolar forceps and loop magnification, dissection is performed just inferior to the zygomatic root to the region of the TMJ (For our early cases we dissected out the origin of the facial nerve and followed out the branches to ensure preservation).
3. Next, an osteotomy of the condylar segment is performed with a curved bunion saw blade placed parallel to the glenoid fossa (inclined slightly inferiorly to avoid penetration into the temporal fossa). A pediatric spine spreader is then placed in this space and opened to stretch the surrounding ligaments. Then, the glenoid fossa is reshaped with a side-cutting bur.



FIGURE 9 Patient images portraying pre-distraction and post-transport distraction appearance. **(A)** Pre-distraction frontal view. **(B)** Post-distraction frontal view. **(C)** Pre-distraction lateral view. **(D)** Post-distraction lateral view.

The pedicled TPF flap is transferred into this space and sutured medially. Gelfoam is placed in the new space to aid hemostasis.

4. Next, through an intraoral approach an osteotomy is performed with a reciprocating saw to separate a posterior mandibular ramus segment from the distal mandible.
5. For fixation a Matthew's Arthroplasty Device (KLS Martin L.P., Jacksonville, Florida) is used (Fig. 12). The cranial fixation arm of the device is placed temporally above the external auditory canal. Self-drilling 2.0 mm×5.0 mm screws are used for fixation to the temporal bone.

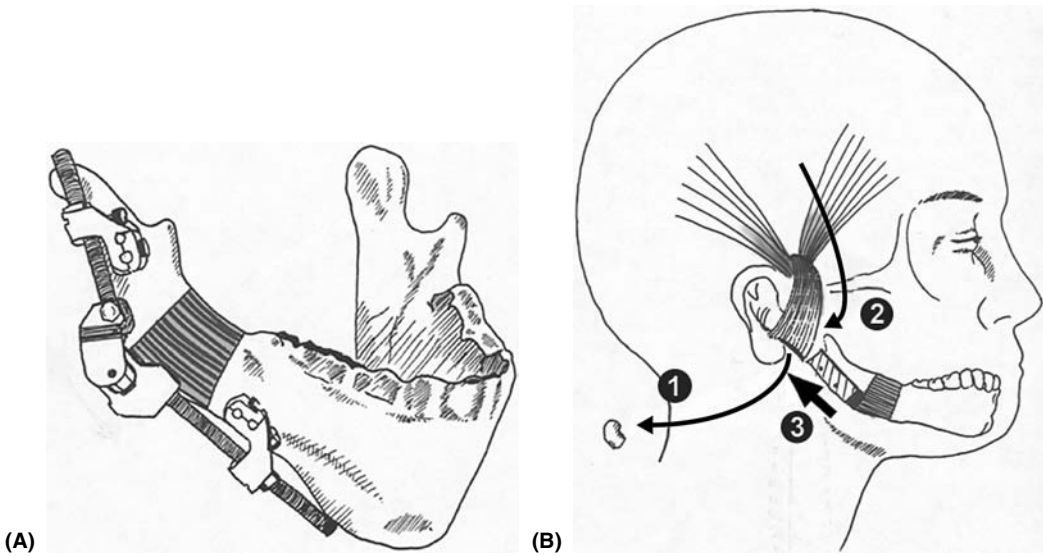


FIGURE 10 Transport distraction osteogenesis device. (A) After initial mandibular lengthening for micrognathia (bony regenerate is indicated by shaded area). (B) In next step, fused condyle is resected, temporoparietal fascial interposition flap is positioned in the glenoid fossa, and the mandibular ramus segment is transported to create a “neocondyle.”
Source: From Refs. 1–3.

If needed, a titanium wedge (provided with the Matthew’s device) is used over the curved (caudad) portion of the temporal bone.

6. Next, the mandibular pins are placed. First, the adjusting screw is turned until the pin anchoring sleeves are over the position of the ascending ramus chosen for pin placement. Then, the pins are placed percutaneously. The adjusting screw is set for 10 to 12 mm of displacement between the condyle and glenoid fossa.
7. Transport distraction is tested under direct vision. Incisions are closed in layers.

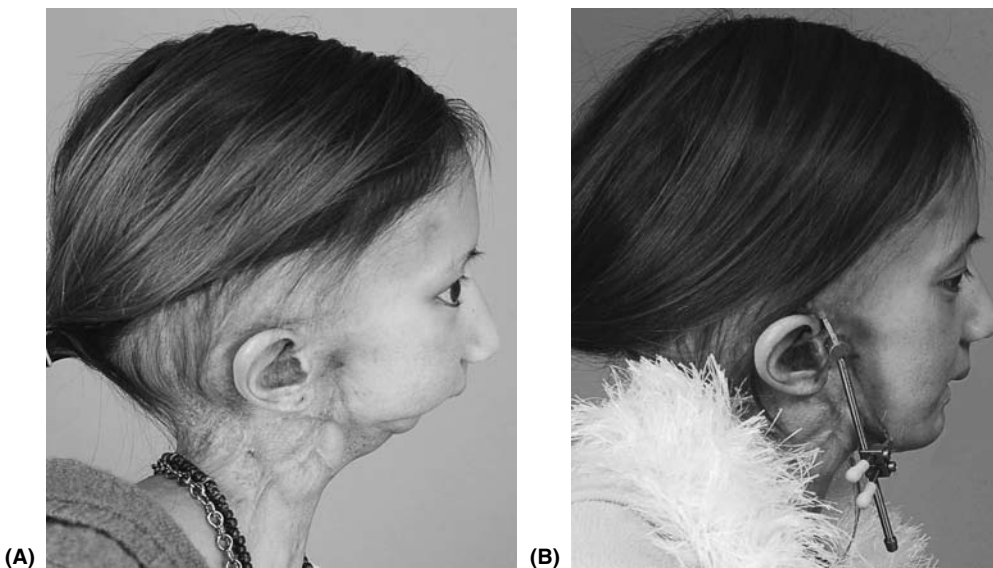


FIGURE 11 Patient images portraying pre-distraction and post-distraction appearance. (A) Pre-distraction lateral view. (B) Post-distraction lateral view with the Matthew’s arthroplasty device in place to allow for maintenance of temporomandibular joint space and movement on the mandible.

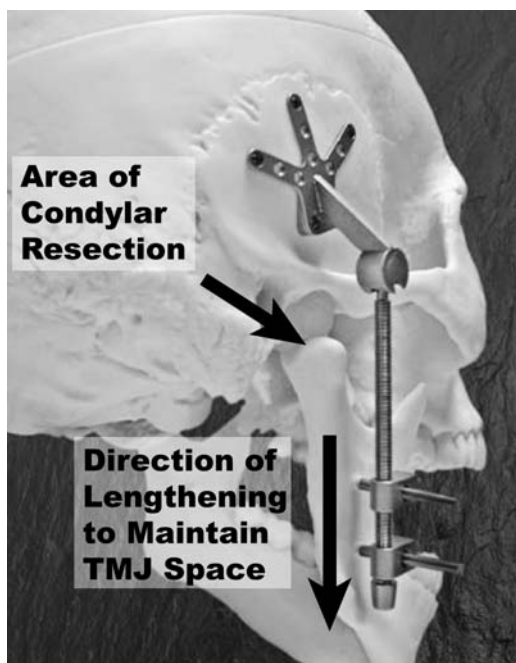


FIGURE 12 Image of Matthew's device. Cranial portion is fixed to temporal bone, and mandibular portion is fixed by two pins to ramus. Distraction directions indicated by arrows.

Transport distraction is begun on day 1 postoperatively at 1 mm per day and consolidation time is continued for three months. Mandibular movement with a Therabite (Atos Medical, Horby, Sweden) is begun as soon as patients have healed their intraoral incisions. The therabite protocol involves three repetitions of 10 jaw stretching exercises four or more times per day. Patients are instructed to chart out daily progress by measuring incisor opening. This therapeutic protocol is continued for at least six months. In addition, patients return to the operation room for mandibular stretching under anesthesia and paralysis, as a routine, every eight weeks for the first six months. After the first six months, therapy and the stretching regimen are continued on a case-by-case basis.

DENTOALVEOLAR DISTRACTION

Distraction osteogenesis of a hypoplastic dentoalveolar segment from lack of dentition, trauma or a congenital deformity has increasingly become a more common method for treatment. In the edentulous patient augmentation of the dentoalveolar segment may be used in preparation for osteointegrated implants. In addition, small defects between alveolar segments, as in clefts, have been successfully treated with translocation of tooth borne bone segments across the defect.

With orthodontic assistance, both vertical and horizontal distraction movements may be used to correct the deficient alveolus. If carefully planned, even alveolar segments containing only one or two teeth may be moved without vascular compromise or injury to tooth roots. In correcting dental malocclusions distraction osteogenesis has become a more important tool for the orthodontist, oral surgeon, and plastic surgeon.

GENIOPLASTY DISTRACTION

An osseous genioplasty is effective in correcting microgenia, vertical facial discrepancies or chin asymmetries. A distraction genioplasty is a two-staged procedure that is only necessary when the soft tissue is tight and the relapse is high. This distraction genioplasty procedure has been used in Treacher-Collins and Nager syndrome patients with a tight soft-tissue envelope

and an existing class I occlusion. For correction of upper airway obstruction genioplasty distraction has been used with a hyoid facial sling advancement procedure.

The hyoid advancement optimizes epiglottal position (since the hyoid has direct ligamentous attachments to the epiglottis). The genioplasty distraction offers a decrease relapse in Treacher–Collins and Nager syndrome patients compared to the traditional acute genioplasty advancement because of the gradual lengthening of the strong genioglossus and geniohyoid muscles which pull in inferior–posterior direction (Figs. 13 and 14).

Genioplasty Distraction Operative Technique

1. Under general anesthesia, a skin incision is made in the submental skin fold and subcutaneous dissection is done down to the hyoid bone.
2. A single hook is used to retract the hyoid toward the chin and the infrahyoid muscle attachments are released with a right angle and bovie cauterizer.
3. A tensor fascia lata graft measuring 2 cm×8 cm is harvested via a lateral thigh incision. This graft is then separated into two slings (1 cm×8 cm each).
4. Fascial slings are placed around the hyoid in a paramedian fashion and sutured to it with 2-0 prolene sutures.
5. Next, a wire-passing drill is used to make two holes in the anterior-inferior mandible and the fascial slings are secured with 2-0 prolene to the anterior mandible.
6. Then, the submental skin incision is closed in two layers.
7. Horizontal osteotomy of the mandible is performed through a lower anterior gingivobuccal sulcus incision.
8. Subperiosteal dissection is performed to expose the site of the osteotomy. An oscillating saw is used to mark the midline and begin the horizontal osteotomy.
9. Osteotomy is completed with a reciprocating saw. (Care should be taken to stay greater than 5 mm below the mental foramen).
10. A burr hole is made in the mandibular bone in the midline (just above the osteotomy) as a channel for the bar of the distraction device. A genioplasty distraction device (KLS-Martin, Jacksonville, Florida), is then secured to the stable bone and genioplasty segment with 2.0 mm×7.0 mm self-drilling screws.
11. Before closure, the turning arm of the distraction device is brought out percutaneously through a small stab incision through the labiomental fold. The gingivobuccal sulcus incision is then closed.

At the completion of 1 mm per day distraction the activation arm is removed. After three months of consolidation the device is removed and a 2-0 titanium right-angled plate is placed for fixation.

ZYGOMATIC DISTRACTION

Zygomatic deficiencies are seen with Treacher–Collins syndrome and in orbitofacial clefts. Traditionally, cranial bone grafts are used for reconstruction in late childhood because this region is known for bone resorption and such grafts often dissolve over time. Recorrection at skeletal maturity may require regrafting or soft tissue filling. Distraction has been attempted and may offer improvement in treatment; however, with zygomatic distraction a secondary device removal procedure is still required.

CRANIAL VAULT DISTRACTION

Craniosynostosis involves restricted growth perpendicular to the fused suture. Craniosynostosis is not a hypoplasia, similar to the above pathologies treated with distraction. However, distraction has been attempted to gradually advance a frontorbital segment in coronal synostosis or widen the biparietal width in sagittal synostosis.

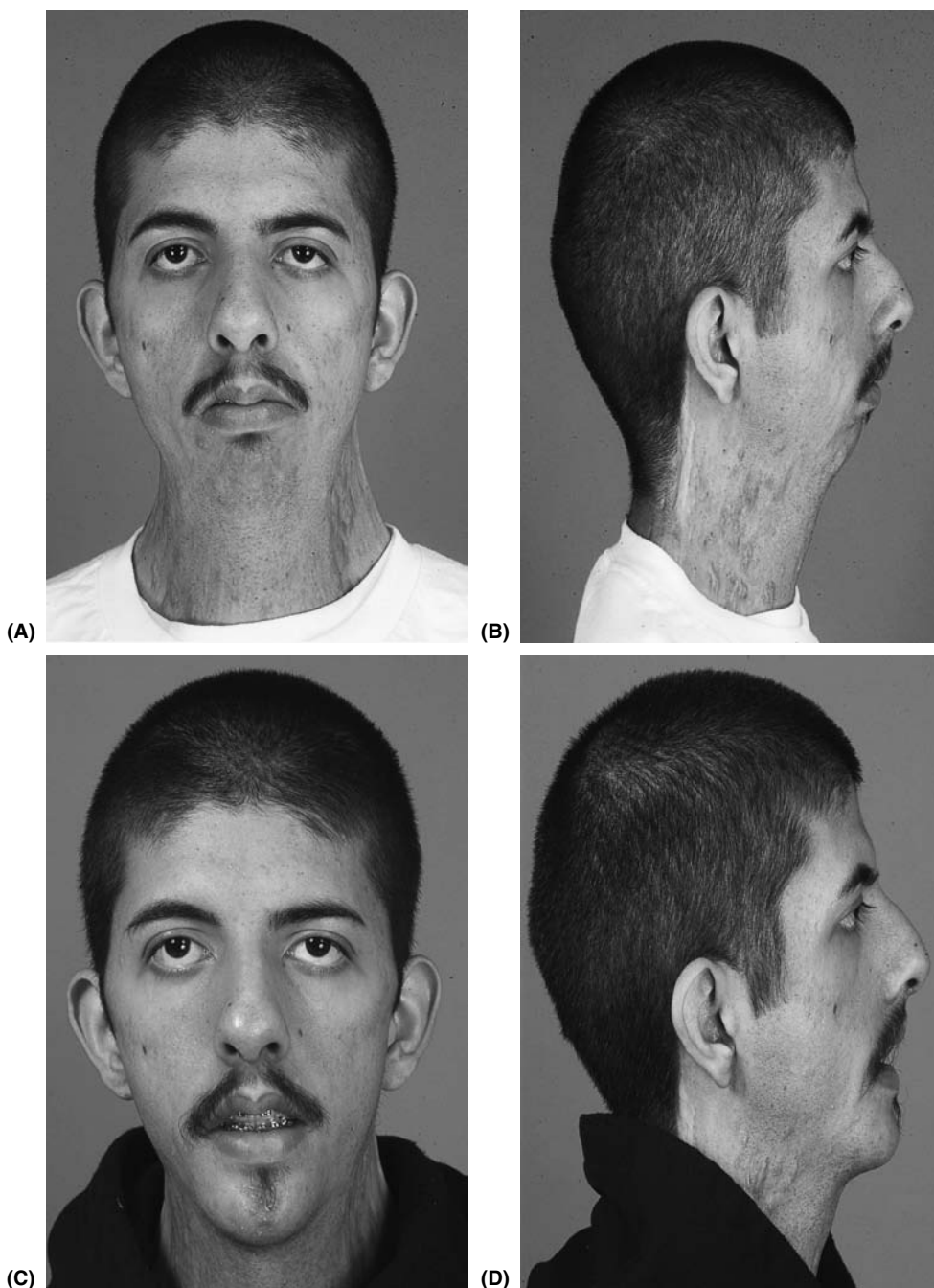


FIGURE 13 Genioplasty distraction. Patient images portraying pre-distraction and post-distraction appearance. (A) Pre-distraction frontal view. (B) Pre-distraction lateral view. (C) Post-distraction frontal view. (D) Post-distraction lateral view.

Some surgeons contest that distraction is not an appropriate treatment tool for craniosynostotic cases. One goal of the process of distraction osteogenesis is the formation of new bone. Yet, a goal in correction of craniosynostosis is to remove the fused suture, provide cranial vault correction and leave space for the normal underlying brain to grow without

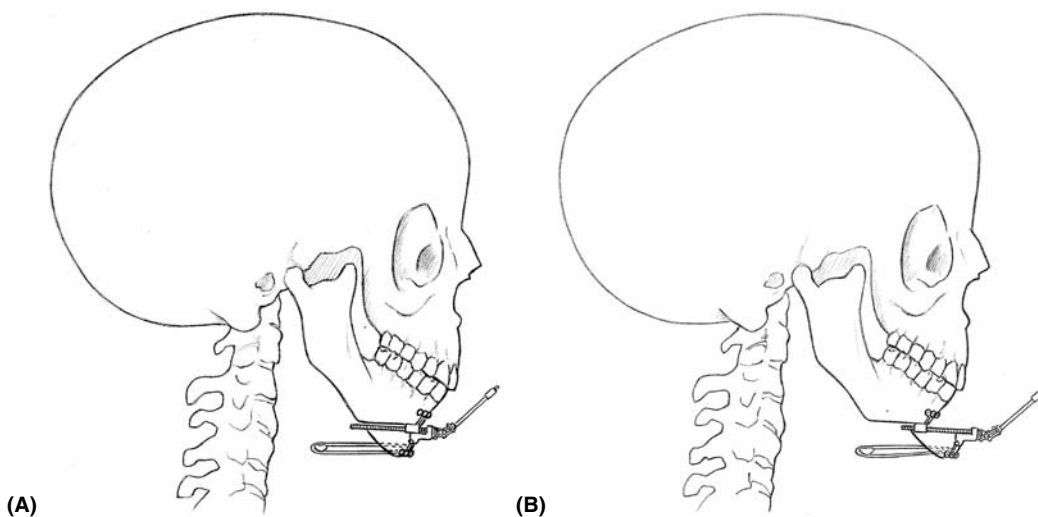


FIGURE 14 Genioplasty distraction technique. (A) Pre-distraction placement of the distractor device is shown with fascial sling about the hyoid. (B) Post-distraction genioplasty with hyoid advancement is shown just prior to distractor arm removal.

restriction. Newly formed distracted bone in the suture area would act similar to a fused suture and restrict subsequent growth. However, there are other modalities, such as “spring-expansion,” which may offer promise as alternative options in the future.

SUMMARY

In Summary, distraction osteogenesis in the craniofacial skeleton is an effective reconstructive option in the growing patient with hypoplasia. In many cases it is superior to traditional grafting procedures with donor site morbidity. The indications for distraction and the instrumentation available will continue to expand. Timing and principles of craniofacial surgery, based on growth and follow-up studies, should be closely followed in this dynamic field.

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Surgical Management of the Temporomandibular Joint

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INTRODUCTION

Of all the skeletal articulations the temporomandibular joint (TMJ) is the most complex. It is two separate synovial joints, a left and right TMJ (bipedalism) that must function in unison in all three anatomic planes and is freely moveable as a single unit. Each has been described as a diarthrodial joint as it has two articulating bony components and functions as a ginglymoid joint because of its initial hinge-like movement. A more accurate description, however, is that each of the joints function as a compound joint and not as a simple hinge movement. For the human mandible, this is a sliding hinge joint implying three bony articulations. The articular disc functions as the “third bone,” supplying true articular surfaces superior and inferior. The TMJ is a double joint composed of an inferiorly positioned hinge joint and a superiorly positioned sliding joint. The disc-condyle complex is a simple hinge joint and the superior joint is designed for sliding movement in any direction, the range being limited by structural restraints. Additionally, the joint position is determined by muscle action until the moment of intercuspation, determined by the occlusion. This is the only joint in the body whose position is not solely determined by resting muscle tone or postural position but determined by an additional variable, the dentition. In many of the craniomaxillofacial conditions, the TMJ may be affected and this chapter focuses on the fundamentals in managing the surgical component of its pathology. Understanding the surgical reconstruction is an understanding of the anatomy of the TMJ.

ANATOMY OF THE TMJ

Structural Elements of the TMJ

The TMJ is the articulation between the condyle of the mandible and the squamous portion of the temporal bone (Fig. 1). Geometrically, the condyle is ovoid in shape with its major axis oriented in the medial lateral direction perpendicular to the ascending ramus of the mandible. The reciprocating skeletal component of the joint is the articular surface of the temporal bone, composed of the concave articular fossa (the glenoid fossa) and the convex articular eminence anteriorly. The glenoid fossa is limited posteriorly by the petrotympanic fissure, medially by the spine of the sphenoid and the foramen spinosum (middle meningeal artery), laterally by the zygomatic tubercle and the postglenoid tubercle, and anteriorly by the descending surface of the articular eminence. Unlike the glenoid fossa, it is the articular eminence that is loaded during function.

The capsule encasing the joint is attached laterally at the zygomatic tubercle, the lateral rim of the glenoid fossa and the postglenoid tubercle. Medially, the capsule is attached to the medial glenoid rim with the spine of the sphenoid, the sphenomandibular ligament and the middle meningeal artery just medial to the attachment. Posteriorly, the capsule attaches to the petrotympanic fissure. Inferiorly, the capsule attaches to the periosteum of the neck of the

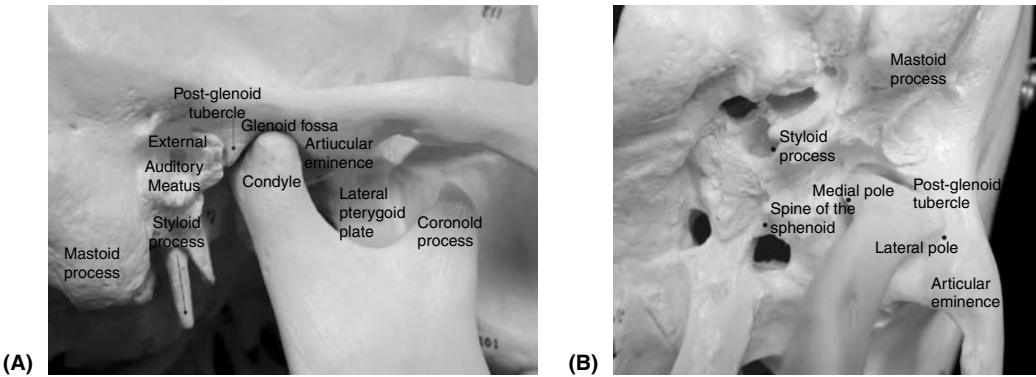


FIGURE 1 Skeletal anatomy of the temporomandibular joint. (A) Lateral view of the articulation of the condyle with the fossa. (B) Infrarossa view of the articulation of the condyle with the skull base.

condyle. A capsule does not exist at the medial half of the anterior aspect of the TMJ and the synovial membrane that lines the superior cavity anterior wall is supported only by loose areolar connective tissue. This lack of an anterior capsule comprises the anatomical “achilles heel” of the TMJ and surgically becomes important in the dissection. The capsule on the medial and lateral wall of the joint is comprised of collagen but the fibers are not under tension. This looseness in the fiber arrangement does not firmly support the joint but allows the condyles to translate forward without tearing the capsule.

Each joint has an intracapsular meniscus (disc), dividing the synovial cavity into superior and inferior compartments, each compartment volumetrically 1 cc or less (Fig. 2). The meniscus is a fibrous, saddle-shaped structure and varies in thickness; a thinner, central intermediate zone separates the thicker rim portions where ligamentous attachments occur. The inferior concavity of the disc precisely fits the convexity of the condyle. Correspondingly, the superior concavity of the disc fits the convexity of the articular eminence. Normally there is no space between these articular surfaces except at the anterior and posterior recesses in the upper compartment and at the anterior and posterior recesses in the lower compartment. These spaces are filled with synovial fluid and allow the condylar movement to occur by the displacement of the fluid between these fluid filled spaces.

The boundaries of these spaces are determined by the disc ligaments. Medially and laterally, the disc is firmly attached to the medial and lateral poles of the condyle by condylodiscal collateral ligaments that are primarily composed of collagen rather than

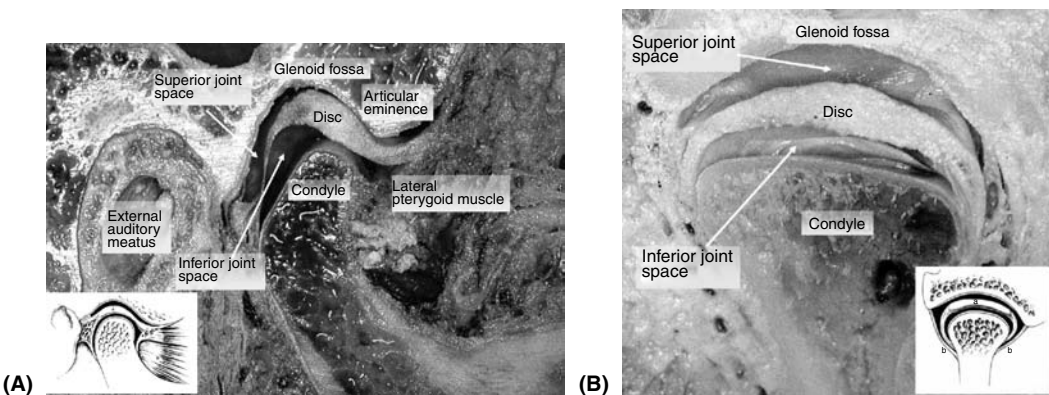


FIGURE 2 (A) Sagittal and (B) coronal sections of the TMJ. *Abbreviation:* TMJ, temporomandibular joint. *Source:* Photos courtesy of Masatoshi Ohnishi.

elastin. Anteriorly and posteriorly, the disc is contiguous with the anterior and posterior attachment tissues called the bilaminar zones or ligament. The bilaminar zone contains loose areolar connective tissue sandwiched by two strata of fibers that is vascular and innervated. It is these bilaminar ligaments that play an important role in allowing the condylar rotation and translation to occur.

The superior stratum of the anterior bilaminar zone attaches to the ascending slope of the articular eminence and the inferior stratum inserts to the anterior aspect of the condyle (Fig. 3). When the condyle is in centric relation, the anterior ligament is relaxed and folded on itself.

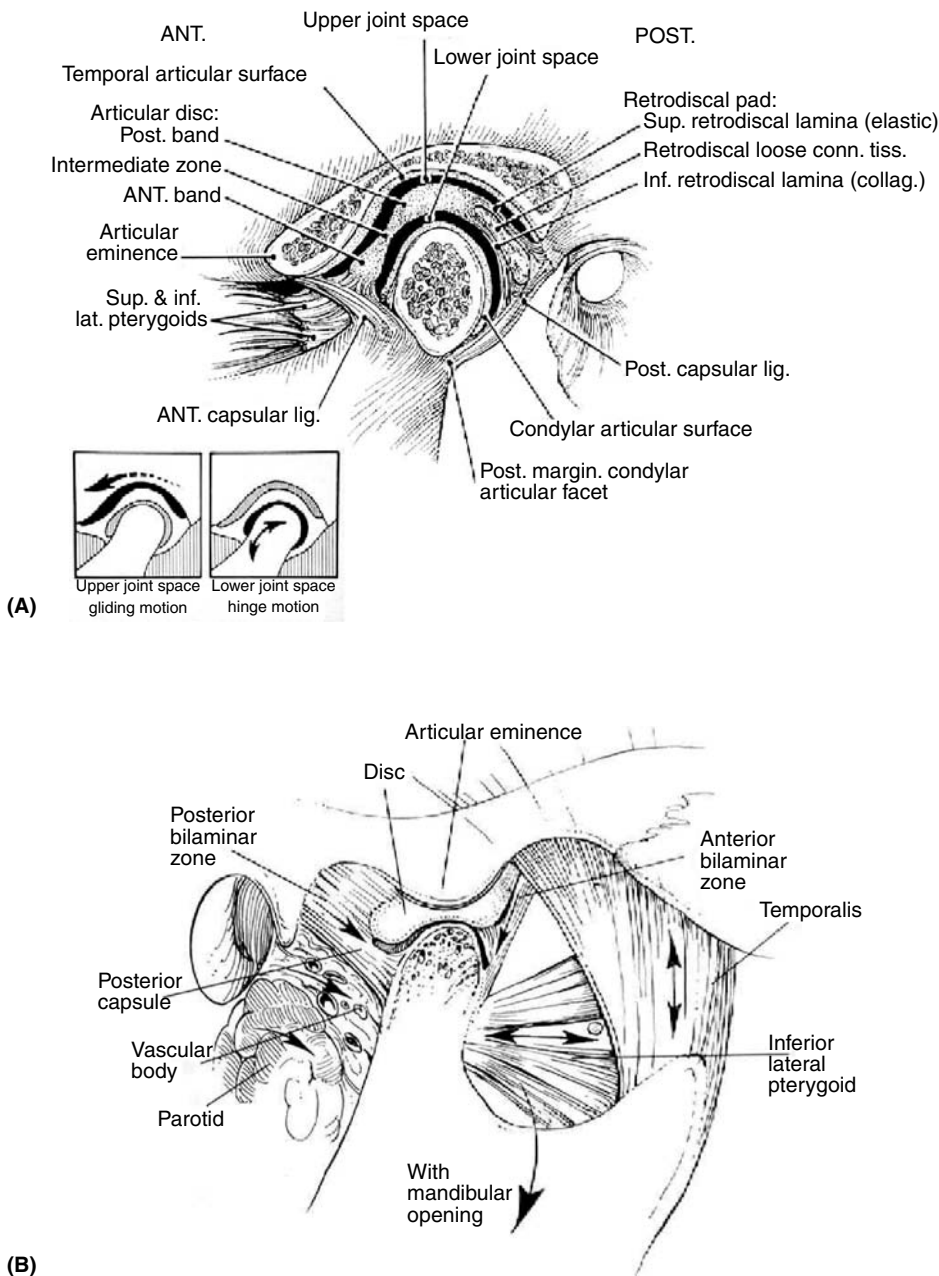


FIGURE 3 Schematic illustration of the intracapsular anatomy of the temporomandibular joint (A) in the closed position and (B) with mandibular opening.

As the condyle rotates, the anterior ligament stretches downward. The superior stratum of the posterior bilaminar zone attaches to the petrotympanic fissure and the inferior stratum to the posterior aspect of the condyle. The superior retrodiscal lamina is comprised of elastin rather than fibrous connective tissue (collagen); and unlike collagen, elastin has a true modulus of elasticity. While the disc is tightly attached to the medial and lateral pole of the condyle, the attachment of the disc to the fossa posteriorly must be elastic to allow the disc to translate forward with the condyle. The inferior stratum of the bilaminar zone is not stretched as the condyle translates forward because the meniscus rotates posteriorly, relieving any tension in this lamina. Thus, functionally the condyle rotates within the encompassing meniscus and together they translate against the articular eminence. This results in less movement and friction between the osseous components of the articulation.

Innervation of the TMJ

The TMJ is innervated by the mandibular branch of the trigeminal nerve (V_3): primarily the auriculotemporal nerve branch with some contributions from the masseteric and deep temporal nerves. The capsule, the disc ligaments, and the synovium are innervated. However, the disc itself along with the fibrous covering of the articular surfaces are not innervated. Additionally, within the TMJ capsule are three mechanoreceptors: the pacinian corpuscles, the Golgi tendon organs, and the Ruffini receptors.

Vascular Supply of the TMJ

The superficial temporal artery, the internal maxillary artery and the middle meningeal artery provide the predominant blood supply to the TMJ. Secondary sources include the deep auricular, ascending pharyngeal and the anterior tympanic vessels. The condyle receives its blood supply through the narrow space of marrow via the inferior alveolar artery and by vessels that directly enter the condylar head.

Mechanics of TMJ Movement

The hinge-gliding movement of the TMJ is controlled by the muscles attached to the mandible. The muscles can be divided into three functional groups: (i) the elevators consisting of the temporalis, masseter, and medial pterygoid muscles; (ii) the depressors which include the digastric, mylohyoid, and geniohyoid muscles; and (iii) the protractors made up of only the lateral pterygoid muscles.

The masseter has two components: the superficial portion whose fibers are oriented in the inferior–posterior direction and the deep portion whose fibers are vertical. When the mandible is open and the powerful bite force of the masseter is applied, its vertical deep portion stabilizes the condyle against the articular eminence. The temporalis can be divided into three distinct components. The anterior portion whose fibers are directed vertically, the middle portion whose fibers are obliquely directed and the posterior portion whose fibers are nearly horizontal. Because of the differential angulations of the fibers, the differential contraction of the temporalis coordinates the closing movement of the mandible achieved by the powerful closure of the masseter. Similarly, the lateral pterygoid in many of the descriptions in the anatomical literature, as having two distinct components: the inferior lateral pterygoid and the superior lateral pterygoid portions. More appropriately, however, these two components should be considered as two separate muscles, as their functions are nearly opposite. The inferior lateral pterygoid originates from the lateral surface of the pterygoid plate, the pyramidal process of the palatine bone and the maxillary tuberosity and inserts into the medial aspect of the condylar neck. When there is bilateral and simultaneous activation of the inferior lateral pterygoid muscles, the condyles are pulled down the slope of the articular eminence and the mandible protrudes. When it activates with the mandibular depressors, the mandible opens. In contrast, unilateral contraction creates a medial movement of the ipsilateral condyle with a resultant movement of the mandible to the contralateral side. The superior lateral pterygoid, in distinction to the inferior lateral pterygoid, is inactive during mandibular

opening, but activates with the elevators. In particular it activates when the mandible closes against resistance as in clenching and chewing. The superior lateral pterygoid originates from the upper third of the lateral pterygoid plate and while majority of the fibers inserts into the medial aspect of the condylar neck, 15% to 40% of the fibers have been described to insert into the disc. This insertion into the disc may play a role during the normal medial rotation that occurs with chewing and that spasm can affect disc position.

Thus, various combinations of the mandibular muscles execute the different movements of the mandible. No muscle acts singly to achieve mandibular movement. A particular muscle may act synergistically with differing muscles at different time points while simultaneously opposing muscles are also act to control the mandibular arc. Additionally, an individual muscle may have portions of it that act differentially. The mandibular movements take place within certain three dimensional limits. The mandible can move laterally 10 mm, protrude approximately 9 mm and retract by approximately 1 mm and open approximately between 45 and 55 mm. During the initial 20 to 25 mm of stomal opening a pure rotation occurs through the condylar axis. To achieve the remaining stomal opening of 45 to 55 mm the condyle must translate forward (Fig. 4). The opening occurs by gravity and relaxation of the elevators. Extensive opening then requires the inferior heads of the lateral pterygoid to apply protracting forces on the condyles and disc unit with simultaneous active depressors acting on the chin and the body of the mandible. Closing movement occurs by relaxation of the opening muscle groups and contraction of the elevators once the disc condyle unit is within the eminence.

Adaptive Remodeling

The articulating surface of the condyle is covered not by hyaline cartilage as it is in the other joints but by an avascular fibroelastic tissue containing chondrocytes that vary with age. In the immature condyle, the deepest layer of this fibrocartilage covering is rich in undifferentiated cells, the reserve layer. Between the reserve layer and the subchondral bone hyaline cartilage

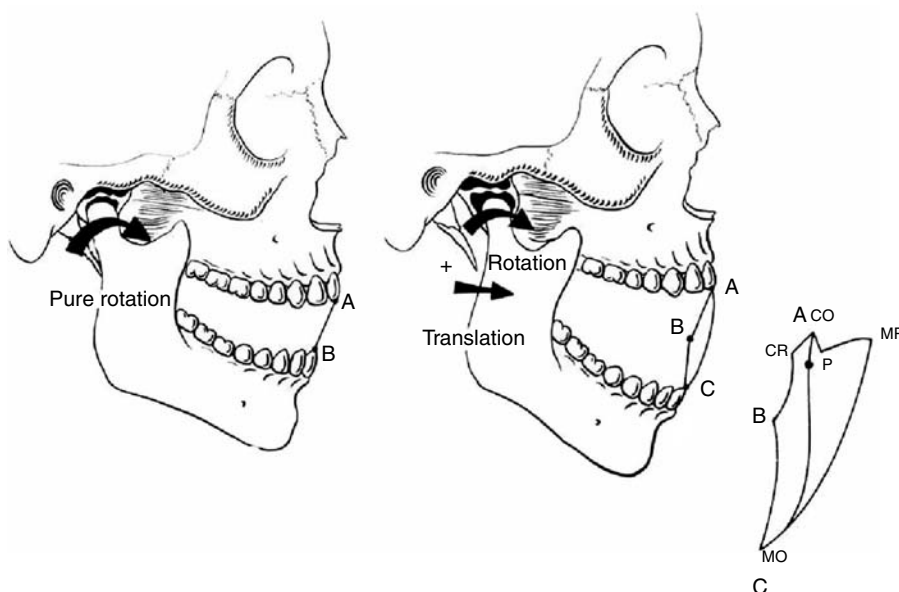


FIGURE 4 Range of motion of the temporomandibular joint in the sagittal plane. As the mandible opens, there is an initial rotation from point A to point B with the center of axis within the condylar head. With further opening the mandible translates from point B to point C and the center of axis moves inferiorly to within the ramal body. The border movements cycles from CO to MO to MP and to CR. *Abbreviations:* CO, centric occlusion; CR, centric relation; MO, mandibular maximal opening; MP, Mandibular protrusion.

does exist, but in the young condyle composed of layers of actively hypertrophying and degenerating chondrocytes. In the presence of cartilage, the immature condyle is able to adapt to loading. However with aging, the condyle becomes calcified, trauma with a change in loading pattern leads to degenerative joint disease (DJD) instead of a hyperplastic remodeling response.

TMJ PATHOLOGY

Each of the structural elements of the TMJ can be affected by congenital, developmental and acquired conditions. The following section briefly discusses the various conditions some of which not specifically related to the TMJ alone [such as hemifacial microsomia (HFM)] are discussed in further detail in other chapters.

TMJ Disorders

A significant percentage of the general population is afflicted with TMJ dysfunction. Depending on the population studied, the percentage is somewhere between 20% and 40% with a predominant female to male ratio of 3:1 to 9:1, afflicting primarily the young to the middle age group. Patients with symptoms specific to the TMJ were present with clicking, popping, locking, grinding, crepitus, and pain. The history should be as detailed as possible and the physical examination of the TMJ systematic. Frequently obtaining the temporal sequence of the symptoms with recall of inciting causes and various tried treatments from the patients at the initial consultation is difficult; and thus, requires a second consultation with the patient keeping a journal and completion of a detailed questionnaire in the interval. The history typically begins in the distant past with severe pain first noticed on yawning, dental procedure or an incidental trauma. The pain and trismus resolve. The episode is frequently forgotten. Sometime later a painless click of the jaw is noted when eating and again an episode of pain is noted on yawning or similar extensive mouth opening. But it occurs infrequently enough to not seek formal evaluation, until sometime later joint stiffness and pain are noted, usually on waking, and the click now occurs more frequently with eating. Pain becomes more consistent aggravated by jaw clenching and worse on awakening. At some point it may progress to difficulty in opening the mouth until a manipulation of the jaw allows it to overcome a "stop." Deviation of the jaw may be noted by the patient with opening of the mouth because of the inhibited forward motion of the condyle on the affected side. At some point it may progress to where the patient is unable to "unlock" the jaw and stomal opening is limited to rotation only. If unresolved, sleep becomes restless, eating difficult and the pain now becomes the focus affecting normal daily life.

Examination in the acute phase will demonstrate a well localized tender joint at the lateral pole of the condyle with severely restricted stomal opening. Masticatory myalgias may be present with tenderness overlying the masseter, the medial pterygoid and temporalis muscles. In the non-acute phase, there is no pain and tenderness of the joint, but there is a pronounced click. Examination should include recording the range of motion (ROM) of the mandible (maximal incisal opening, lateral excursion and protrusion) noting any deviation from the midline, auscultation of the joint and noting the timing of TMJ sounds in the opening-closing cycle, palpation of the condyle for subluxation or dislocation of the joint with mandibular excursion, recording the areas of localized joint and muscle tenderness, assessing the occlusion and the dentofacial skeletal relation.

The temporomandibular joint disorders (TMJD) in actuality are a *collection* of disorders that are not clearly understood with many overlapping features and a myriad of therapeutic options. It is not a single entity. Conditions typically grouped within TMJD include internal joint derangement (IJD), DJD, and myofacial pain (MFP).

Internal TMJ Derangement

IJD occurs as a disruption of the intracapsular components of the TMJ where there is an abnormal relation of the disc relative to the condyle, the glenoid fossa and the articular

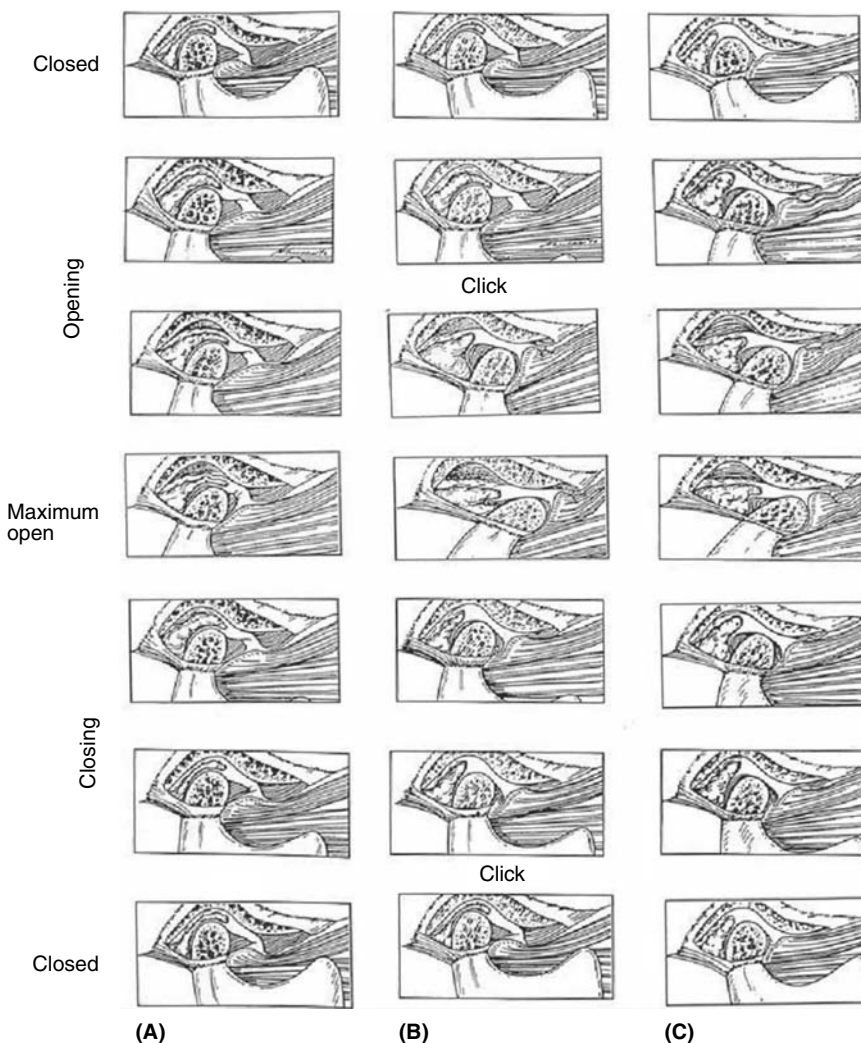


FIGURE 5 (A) Disc dislocation without reduction. The condyle is unable to assume a normal relationship with the disc as in B. The disc moves ahead of the condyle and restricts translational opening of the mandible. The condyle never “captures” the disc. (B) Disc dislocation with reduction. The condyle passes over the dislocated disc onto the intermediate area during opening (recaptures the disc). This reduces the disc and there is a click. Remaining opening and closing is normal until the last portion when the disc is again dislocated and a click occurs. Note that the click is asymmetric in the cycle. Temporomandibular joint function during the full range of opening and closing. (C) Normal disc function. The disc is rotated posteriorly on the condyle as it is translated out of the fossa.

eminence (Fig. 5). The disc is frequently displaced anteriorly resulting in abnormal mandibular function. The severity may range from intermittent occurrence to complete displacement with disc perforation (Table 1) and surrounding tissue reaction with resulting synovitis, scarring and adhesions, fibrocartilaginous metaplasia and calcification. Symptomatically, the patients are present with pain that specifically occurs with loading of the TMJ and is localized to the TMJ. Additionally, joint clicking or popping if present must also be coincident with the pain. At the time of the symptoms, limitation of ROM must also be present. Examination should confirm reciprocal clicking with opening and closing at different condylar positions (stages I and II), tenderness to palpation, deviation with opening without significant translation (stages III and IV), and crepitus (stage V). If this constellation of symptoms and signs is not closely coupled, then it is unlikely that IJD is the likely cause of the pain.

TABLE 1 Wilkes Classification of Internal Joint Derangement

Stage	Condition
I	Early reducing disc displacement
II	Later reducing disc displacement
III	Acute/subacute nonreducing disc displacement
IV	Chronic nonreducing disc displacement
V	Chronic nonreducing disc displacement with osteoarthritis

Degenerative Joint Diseases

One must also remember that DJD can equally affect the osseous morphology of the condyle, the articular fossa, and eminence as with any other joint and be a contributing factor to TMJD. Causes include osteoarthritis (OA), juvenile rheumatoid arthritis, rheumatoid arthritis, and ankylosing spondylitis in which the TMJ is involved. Thus, a more general medical assessment of the patient is important in such conditions to place the TMJ component in appropriate context.

Masticatory Myalgias

Masticatory myalgias are frequently included with TMJDs. The belief is that structural abnormalities such as occlusal interferences, malocclusions and malposition of the condyle relative to the fossa or malposition of the internal disc result in muscular dysfunction and pain. The muscles work harder to compensate the structural abnormality and a vicious—hyperactivity circle is set into motion. However, careful review of the literature fails to support this. The matter is further confused by the addition of mental stress tension headaches, regional MFP and fibromyalgias to this grouping of TMJD when no obvious structural abnormality is detected. While displacement of the disc may be accompanied by masticatory myalgia, the converse is not true. It should not be assumed that the disc is displaced if MFP is present alone. There is no clear association between myalgias and structural abnormalities and the search for structural solution requiring surgical intervention should be abandoned in the vast majority of cases.

Malocclusions and TMJD

Similarly, one would expect a causal relationship between dentofacial skeletal malocclusions and TMJ symptoms. Correction of the malocclusion with orthognathic surgery would expect to consistently provide relief of TMJ symptoms by restoring centric occlusion with centric relation. However the literature is not fully supportive. At best the literature suggests there is trend that in a significant number of patients with dentofacial skeletal deformity and TMJ disorders will experience an improvement in their symptoms, but reliably predicting prior to surgery which particular patient will improve is impossible and that in some temporomandibular disorders (TMD) may develop who had none prior to orthognathic surgery. While orthognathic surgery has minimal role in the treatment of IJD, comprehensive treatment may require addressing both components and at least a frank discussion with the patient that IJD is a separate entity. If malocclusion is a contributing factor to TMJD, then it is likely MFP than IJD. A centric relation splint may help alleviate MFP which is of occlusal origin. The corollary is that if splint therapy does not, then the likelihood of orthognathic surgery to relieve MFP is unlikely. Unfortunately, splint therapy as a diagnostic value is not reliably predictive. Even in the absence of symptoms, the prevalence of radiologically diagnosed disc pathology ranges from 21% to 32%. Thus, the indication for orthognathic surgery is malocclusion and not correction of symptomatic TMJ. Finally, what role orthognathic surgery has to prevent future development of TMJ symptoms remains equally unclear.

Structural TMJD

The congenital deformities are not formally categorized within the spectrum of TMJD. However, in many, the mandibular structural abnormality includes TMJ as a component and the patients surprisingly have no TMJ symptoms given the marked involvement

in those that do have joints. Additionally, this would include TMJ ankylosis, dislocations, condylar/subcondylar fractures and tumors. This broad category includes all other TMJ pathology not specifically involving the internal TMJ mechanics of the disc condyle relation discussed in the previous section.

IMAGING THE TMJ

Imaging the TMJ can be difficult. For it to provide diagnostically relevant information it must assess not only the skeletal elements of the joint but also the disc itself. Additionally, it must provide information as the joint dynamically functions in time. Specialized plain radiographic views such as the open and closed mouth transcranial projection are of limited value and orthopantomograph is valuable only as screening radiographs to exclude other pathology. The plain films are normal in up to 85% of the patients with TMJ dysfunction, thus its diagnostic value is questionable. Computed tomography (CT), magnetic resonance imaging (MRI), contrast arthrography and diagnostic arthroscopy are more relevant (Fig. 6).

CT is useful in delineating the skeletal anatomy of the condyle and the zygomatic arch. Data acquired as a spiral CT scan at most centers today (0.5 to 1.0 mm) is extremely fast reducing motion artifact, generating volumetric information that can be reformatted into multiple planes and reconstructed in three dimensions with imaging software. However, the disc cartilage, joint capsule and the surrounding muscles have similar densities and radiographic isolation of the structures is difficult even with adjustment of the window settings and soft tissue algorithms. Additionally, obtaining dynamic information of the joint function (open and closed mouth) requires rescanning which is rarely reproducible for accurate comparison and additional radiation dosage. Thus, the CT is only useful for identifying the skeletal pathology in congenital malformations seen in craniofacial/hemifacial microsomia and related conditions, in acquired conditions as condylar/zygomatic fractures and skeletal tumors. It provides the structural baseline for the MRI scan.

Disc pathology is better noninvasively assessed through MRI scans. MRI relies on the relative proton (H^+) density within the tissue volume and is better suited at assessing soft tissue pathology over skeletal conditions. Imaged primarily in the sagittal plane with surface coils and high resolution technique, the disk is identifiable as a low signal (black) tissue within the higher signal of the synovial fluid and the bilaminar structures above the condyle. The cortex of the condyle is low intensity and surrounds the higher intensity (white) signal of the fat within the marrow. The dynamic positioning of the disk can be artificially obtained with series of static MRIs obtained at various the stomal openings using bite blocks. While magnetic resonance is better suited to diagnosing internal derangement and complements the skeletal information from the CT scan, it falls short of providing true dynamic positioning with normal opening and closing of the TMJ.

While MRI is universally available and is noninvasive, arthrography though invasive provides detail that current MRI technology cannot. Disorders of the disk were well defined by arthrographic studies from two decades ago. However, it requires equipment and staff that is available only in certain centers and is highly dependent on operator skills. The technique involves introducing a small gauge needle into the inferior joint space immediately posterior to the condyle and injecting iodinated contrast under fluoroscopy. The contrast tracks along the posterior, superior and anterior portions of the condyle. The anterior collection of contrast normally has a smooth, teardrop shape. If the meniscus is perforated, contrast flows into both the superior and inferior joint recesses. As the condyle translates anteriorly, the contrast usually empties from the anterior recess and flows posteriorly. When the meniscus is anteriorly displaced, the anterior recess becomes abnormally elongated. Often the displaced meniscus is deformed or buckled, which results in a mass effect against the contrast in the anterior recess. As the condyle translates anteriorly, the mass effect against the anterior recess often increases. When the meniscus reduces, the anterior recess returns to a normal appearance. If the meniscus does not reduce, the anterior recess remains deformed in the fully open mouth position. Not all patients suggestive of disk problems should have arthrography performed, but when surgery is indicated based on clinical grounds it is useful to help exclude those who will not benefit from surgery.

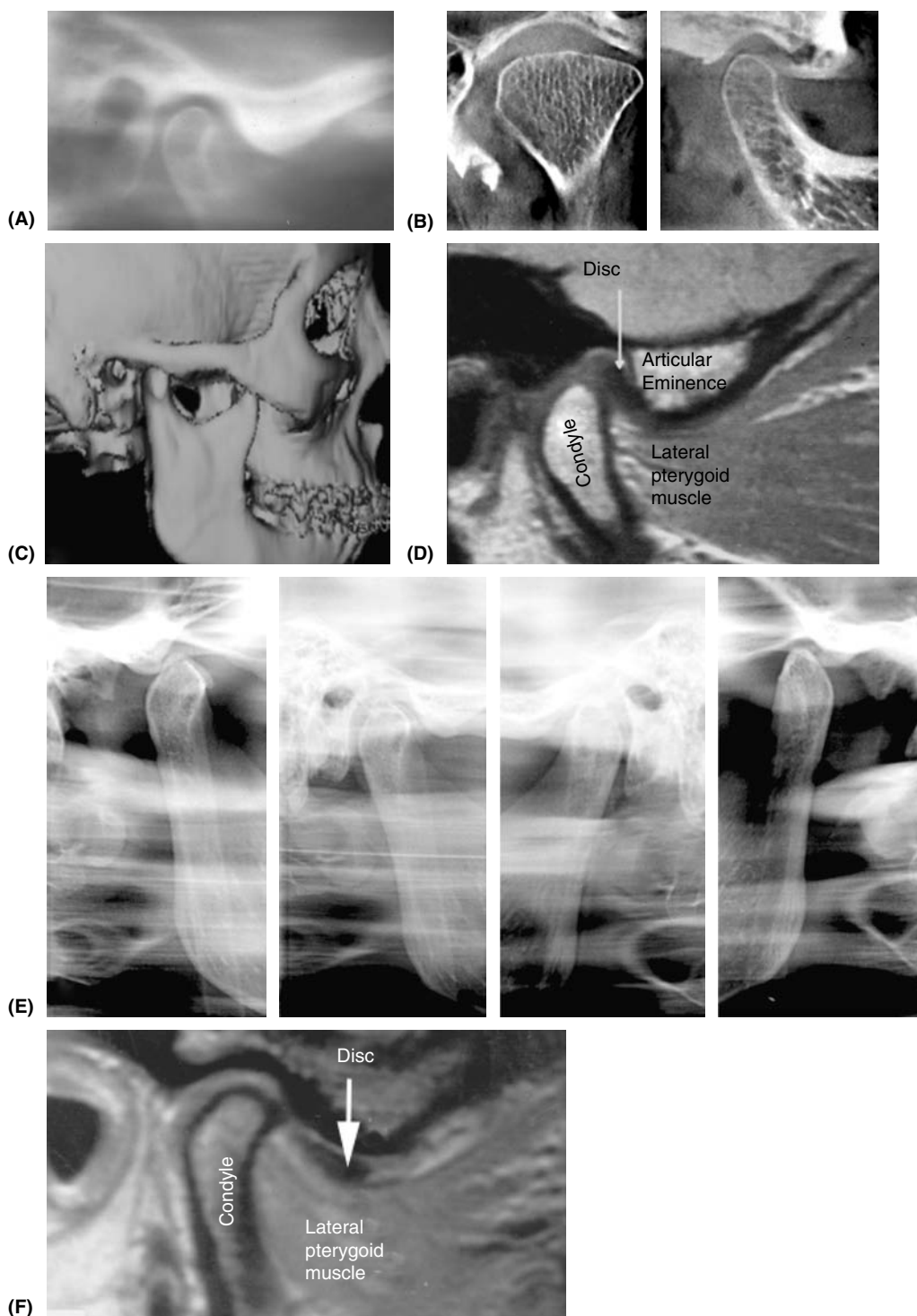


FIGURE 6 Radiographic evaluation of the temporomandibular joint: (A) orthopantomograph; (B) 2-dimensional computed tomography (CT) scan; (C) 3-dimensional CT scan; and (D) magnetic resonance imaging (MRI). Dynamic assessment can be obtained with (E) open and closed views and with (F) MRI imaging that can show disc displacement.

Diagnostic arthroscopy can be used as an alternative as it allows direct visualization of the joint clarifying the clinical diagnosis and simultaneously affords diagnostic biopsies and treatment. Whether it is performed for diagnostic reasons is largely determined by where the above radiographic techniques fail to provide the needed information to determine the course of treatment. The usual indications include internal derangement, OA, tumors and posttraumatic complaints. Detailed discussion on arthroscopic technique is left to authoritative texts solely devoted to the subject.

NONSURGICAL MODALITIES FOR TMJ MANAGEMENT

With the exception of skeletal deformities that include the TMJ and require reconstruction, nonsurgical modalities remain the mainstay for the vast majority of the patients for symptomatic relief (TMJD). However, it should be remembered that these therapies remain an important adjunct to improved outcome in surgical reconstruction (postoperative care in the management of TMJ ankylosis as an example). Treatment include physical therapy and jaw mobilization exercises to restore muscle resting tone, biofeedback therapy to alleviate stress, muscle relaxants and non-steroidal anti-inflammatory agents to reduce muscle spasms and joint inflammation, altering the diet and orthotic splint appliances to decrease loading of the TMJ and “de-program” the muscles of mastication. Surgical intervention for TMJD is considered only with failure of an adequate trial of conservative nonsurgical therapy and these are described in the following section.

SURGICAL APPROACHES TO THE TMJ REGION

The ideal approach to the TMJ, the base of the zygomatic arch and the condylar neck must be based on clear anatomic landmarks, protection of facial and auriculotemporal nerves, avoid injury to the external auditory canal (EAC), good visibility of the operative region in a bloodless field, rapidly executed with minimal operative time, uncomplicated repair of the approach, good appearance of the surgical scar and easily teachable. Multiple surgical approaches with various modifications to access the TMJ have been described and these can be grouped in the following categories: (i) the preauricular approach (endaural), (ii) the postauricular approach, (iii) the submandibular approach (Risdon), (iv) the intraoral approach, and (v) the arthroscopic approach. Thus, there is no single ideal approach. Each of the above approaches has its indications, advantages and disadvantages, and thus the surgeon needs to be familiar with all approaches. Common to all open procedures is the use of general anesthesia without a long acting paralytic agent for intraoperative monitoring of the facial nerve function, the use of a nerve stimulator at selective points in the exposure, bipolar electrocautery and a local anesthetic containing a vasoconstrictor to reduce bleeding and systemic perioperative course of intravenous antibiotics.

Preauricular Approach

The preauricular incision is made closely following the ear crease with either a pretragal or retrotragal variation as with aesthetic surgical procedures. A temporal extension superiorly as described by Rowe, Obwegeser and others allows further exposure to the limited operative field. The temporal extension reduces the chance of facial nerve injury due to flap retraction and the limitations of “keyhole” surgery. With surgical experience, the temporal component of the incision can be limited. Once the incision is made, the temporalis fascia superiorly should be clearly identified and the dissection plane is just deep to the superficial temporoparietal facial to preserve the facial nerve branch to the frontalis muscle. This should be elevated as part of a composite skin flap as the dissection is carried anteriorly. The superficial temporal vessels can be avoided by dissecting close to the cartilage of the pinna. Just anterior to the tragus, the dissection should follow closely the cartilaginous surface of the EAC. In the course of the exposure it is essential to closely follow the nonlinear course of EAC, otherwise the canal may be transected, the tympanum exposed, and the bony EAC mistaken for the glenoid fossa. This surgical plane is an avascular plane between the EAC and the glenoid lobe of the parotid gland.

This approach leads directly to the glenoid tubercle. This is a key landmark. The tympanic membrane attaches to the EAC at the level of the bony canal, and the dissection along the anterior aspect of the EAC cartilage must stop at the postglenoid tubercle. At this point the base of the zygomatic arch can be palpated and identified clearly by clearing off the tissue to the suprapariosteal plane. This should be at the same depth of plane as the temporalis fascia approached from the superior aspect of the incision and the postglenoid tubercle.

Exposure of the TMJ capsule is now guided by following the surface of the zygomatic arch. The temporalis fascia is divided approximately at a 45° angle to the axis of the zygoma beginning at the root of the zygoma and extending for approximately 2 cm in the anterior superior direction. At this point exposure of the planes is easier with a periosteal elevator and with only selective use of fine scissors. The dissection continues anteriorly between the superficial and the deep layers of the temporalis fascia along the suprapariosteal plane of the zygoma. A number of vessels will be encountered and these should be selectively cauterized to allow exposure deep to these vessels. The dissection proceeds anteriorly along the zygomatic arch until the articular tubercle is identified. With the periosteal elevator, the exposure is now continued at the suprapariosteal level inferiorly separating the capsule from the overlying tissues to the neck of the condyle. The exposure to the TMJ region is now complete: the temporalis muscle above, the base of the zygoma with the overlying periosteum from the postglenoid tubercle to the articular tubercle, the lateral aspect of the TMJ capsule and the neck of the condyle.

Postauricular Approach

When greater exposure is desired, a postauricular approach can be considered. The skin incision should be placed along the auricular crease with the ear in the flexed position. The incision is deepened to the mastoid fascia by transecting the posterior auricular muscle. The mastoid fascia is contiguous with the temporalis fascia and the dissection superiorly can be continued on this plane anterior to the ear canal. The posterior aspect of the EAC can be further defined on the mastoid fascia plane. It should be noted that the posterior wall of the EAC is not cartilaginous and an inadvertent perforation can occur. Next a subcutaneous tunnel below the canal is bluntly developed with scissors. A broad blade (#11 or #10) is then introduced into this subcutaneous tunnel with its cutting edge superiorly directed. The helical rim of the ear is then retracted posteriorly and the knife is smoothly (without a sawing motion) brought upward completely transecting the pinna from the EAC. The blade must be visualized as it transects the pinna, controlling the depth. The transaction should not be so deep that reapproximating the pinna to the canal can be easily accomplished. The external ear now anteriorly pedicled on a broad base is retracted forward exposing the stump of the EAC. The connective tissue is dissected off of the cartilaginous EAC to the level of the temporalis fascia. The canal can be temporarily sutured closed to protect it. The posterior edge of the parotid gland is bluntly dissected free and reflected anteriorly. At the level of the temporalis fascia the base of the zygomatic arch to the articular eminence can be palpated. The remainder of the exposure of the TMJ capsule is as described in the preauricular approach beginning with incising the superficial layer of the temporalis fascia and continuing the dissection anteriorly deep to this layer. The posterior approach allows retraction of the EAC stump and a better view of the posterior aspect of the joint than the preauricular approach.

While greater exposure of the TMJ can be achieved with the postauricular approach, the disadvantage is the longer operative time, the infrequent risks of EAC meatal stenosis and infection and temporary hypoesthesia of the pinna for a period of several months. Attention to detail by transecting pinna within the outer third of the EAC and meticulous closure would minimize the risk of stenosis. Similarly, detailed preoperative cleansing of the operative field including the canal and the use of systemic antibiotics has significantly reduced the risk of infection. It is an excellent approach with minimal morbidity; however, its universal application is primarily limited by most surgeon's level of comfort in transecting the external ear.

Sub-Mandibular Approach

In cases of bony ankylosis, fixation of condylar fractures and costochondral reconstruction of the TMJ, the preauricular approach is insufficient and additionally a counter submandibular approach is also needed to expose the angle and the posterior border of the ascending ramus toward the condylar neck. It is important to remember that simultaneous exposure from both approaches must leave an intact bridge of soft tissue from the bony external auditory meatus to a variable distance inferior because of the main trunk of the facial nerve. Though Risdon's name is associated with the submandibular approach, Risdon did not describe it in sufficient detail and modern variations take into account anatomical studies describing the course of the mandibular and cervical branches of the facial nerve. In general, for the adult, the skin incision should be placed approximately 2 to 3 cm below the mandible border to minimize the risks to these nerves. However, in children and in particular children with hemifacial microsomia, no clear guidelines can be given for placement of the submandibular incision. The surgeon can rely only on careful elevation of the planes with loupe magnification and the use of a nerve stimulator. The incision is made through the skin, the subcutaneous fat and the platysma at the level of the cervical fascia. The dissection is then continued superiorly with the nerves protected within the composite skin flap. Anteriorly, the facial artery and vein may be encountered and require selective ligation to allow adequate exposure. The inferior border of the mandible and angle can be palpated with a periosteal elevator. The masseter muscle is then divided along the inferior border and the periosteum incised. The mandibular angle and posterior border of the ascending ramus are exposed in the subperiosteal plane. Visualization of the condylar neck can be further facilitated by engaging a channel type retractor within the sigmoid notch the working operative field is within a long "tunnel."

Intraoral Approach

In cases requiring subcondylar osteotomy and hyperplastic lesions of the condyle, the intraoral approach is ideal. With this approach, the facial nerve and the EAC are not placed in jeopardy. However, the lingual nerve, the inferior alveolar neurovascular bundle and the internal maxillary artery are at risk. An intraoral mucosal incision is made by palpating the ascending ramus. The anterior border of the ramus is exposed in the subperiosteal plane along the coronoid process. The attachments of the temporalis muscle are removed from both the medial and lateral aspects. A narrow width periosteal elevator is used to create "tunnels" on either side well above the inferior alveolar neurovascular bundles. Once the posterior border of the condylar neck is identified, Obwegeser retractors can be placed from both the medial and lateral aspects. Adequate exposure of the condylar neck may require a formal coronoidectomy.

Arthroscopic Approach

When the pathology is solely within the joint capsule itself, the arthroscopy approach appears ideal in that it allows direct visualization of pathology and limited procedures with minimal morbidity to the patient. However, a limitation is that only the upper joint space can be examined. The scope is introduced into the superior joint space either via anterosuperolateral or posterosuperolateral or endaural approach. This allows examination of the posterior joint space, the intermediate zone and the eminence. The mobility of the disc can be examined. Areas of adhesions, perforations, synovitis and chondromalacia can be detected. Procedures that can be addressed through arthroscopy include lysis of adhesions that prevent disk movement during condylar translation, disk repositioning in patients with chronic locking of the TMJ, lavage of microcartilaginous and inflammatory products and in patients with OA, synovectomy in patients with arthritides and treatment of TMJ hypermobility by scarring the posterior disk attachment. While the increasing complexity of the procedures that can be carried out in advance with the specialized instrumentation and the surgeon's experience, the long-term outcome compared to open arthrotomy remains proven. Detailed discussion on arthroscopic technique is left to authoritative texts solely devoted to the subject.

SURGICAL MANAGEMENT OF TMJ PATHOLOGY

There are a variety of conditions that range from impaired hypoplastic under development to over development excessive growth conditions that specifically alter the normal anatomy and subsequent development of the TMJ. Additionally, within this category are the acquired fractures, dislocations and ankylosis. These are primarily structural problems. Surgical management of internal capsular condylar disc mechanism is considered separately in the following section.

HFM and Related Congenital Deformities

In congenital conditions as craniofacial/hemifacial microsomia spectrum and Treacher-Collins/Nager syndrome, the ascending ramus is variably affected from completely absent TMJ structures (condyle, glenoid fossa, and disk) to where all the structures are present but diminutive in size (Fig. 7) Reconstruction depends on delineating the anatomic defect and staging the reconstruction with distraction osteogenesis in Pruzansky Class II conditions to constructing a pseudo-TMJ with costochondral graft reconstruction in Pruzansky Class III along with staged distraction and conventional orthognathic surgical techniques. Management is described in Chapters 16 and 17.

TMJ Hypermobility and Recurrent Condylar Dislocation

Acute dislocation of the condylar head occurs when it extends beyond the articular eminence. Frequently spontaneous reduction occurs. When it does not then it can be manually reduced. Intravenous sedation and muscle relaxants are given before the manipulation. The posterior mandible is grasped by placing thumbs intral-oral with fingers wrapping extral-oral around the body of the mandible. Pressure is applied downward and anterior in a counter-clockwise direction to drop the condyle below the eminence, this then is followed by posterior pressure to reseat it



FIGURE 7 Clinical case illustrating reconstruction of hemifacial microsomia. Coventional orthognathic surgery combined with costocartilage rib grafting to reconstruct the absent mandibular ramus and temporomandibular joint.

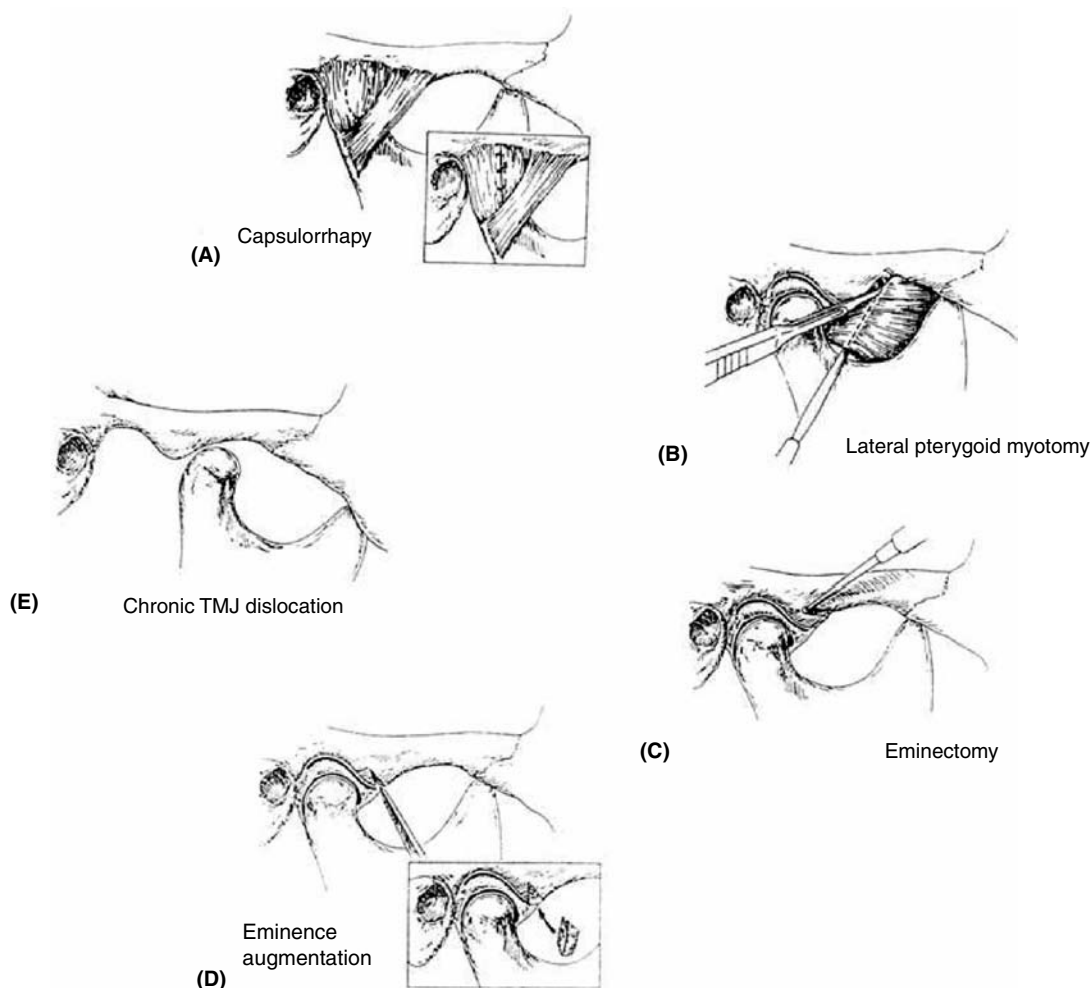


FIGURE 8 Surgical option in recurrent condylar dislocation: (A) capsulorrhaphy, (B) lateral pterygoid myotomy, (C) eminectomy, (D) eminence augmentation, and (E) chronic TMJ dislocation. *Abbreviation:* TMJ, temporomandibular joint.

within the fossa. If this fails then a block of the mandibular branch of the trigeminal nerve should be considered. A spinal needle is directed through the sigmoid notch until the needle is blocked by the lateral pterygoid plate. It is then redirected posteriorly by 1 cm. The local anesthetic is then infiltrated into the lateral pterygoid muscle. If this fails, then manipulation under general anesthesia is considered. Once the condyle is reduced, the maxillary-mandibular opening should be restricted for a period of several weeks to allow capsular healing and prevent spasm. Surgery should only be considered with chronic recurrent episodes of condylar dislocation. Options that have been described in the literature include augmentation of the eminence with autogenous bone graft, removal of the eminence (eminectomy), external pterygoid myotomy and capsulorrhaphy (Fig. 8). Of these, eminectomy is surgically straight forward and effectively prevents locking (Fig. 9). It is the treatment of choice.

TMJ Hypomobility and Ankylosis

Hypomobility or ankylosis may be caused by either intracapsular or extracapsular pathology. The causes of ankylosis include trauma, intracapsular hemarthrosis, septic arthritis of the TMJ originating from suppurative infection of the middle ear, arthritides of various origins and

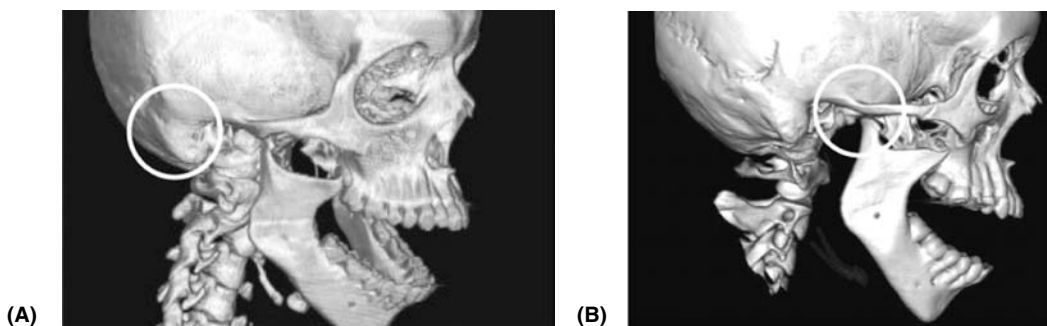


FIGURE 9 Computed tomography scan demonstrating (A) before and (B) after eminectomy in a patient with chronic temporomandibular joint dislocation and locking. Procedure relieved symptoms.

congenital conditions. Regardless of the etiology, the process results in progressive destruction of the intracapsular elements from fibrous adhesions to frank bony union. Patients do have some mobility due to the flexibility of the mandible and the cranial sutures. With bony ankylosis, interincisal opening is 5 mm or less and true protrusive function is absent. When the ankylosis occurs before skeletal maturity, a typical Class II dentofacial skeletal deformity occurs. Routine dentofacial skeletal films and CT scan are needed for surgical management. While the extent of the TMJ pathology is obvious, the coronoid process should be assessed for enlargement and impingement as cause.

When ankylosis occurs, enbloc resection is required with management ranging from a gap arthroplasty to restoring the TMJ using costochondral grafts in children with prosthetic implants reserved only for adults though, even in adults autogenous reconstruction is preferred. Results from ankylosis surgery can be disappointing, but failure can be lessened with aggressive removal of bone, stripping of muscles, coronoidectomy, interpositional tissue to prevent reankylosis, and long-term postoperative physical therapy (Figs. 10 and 11).

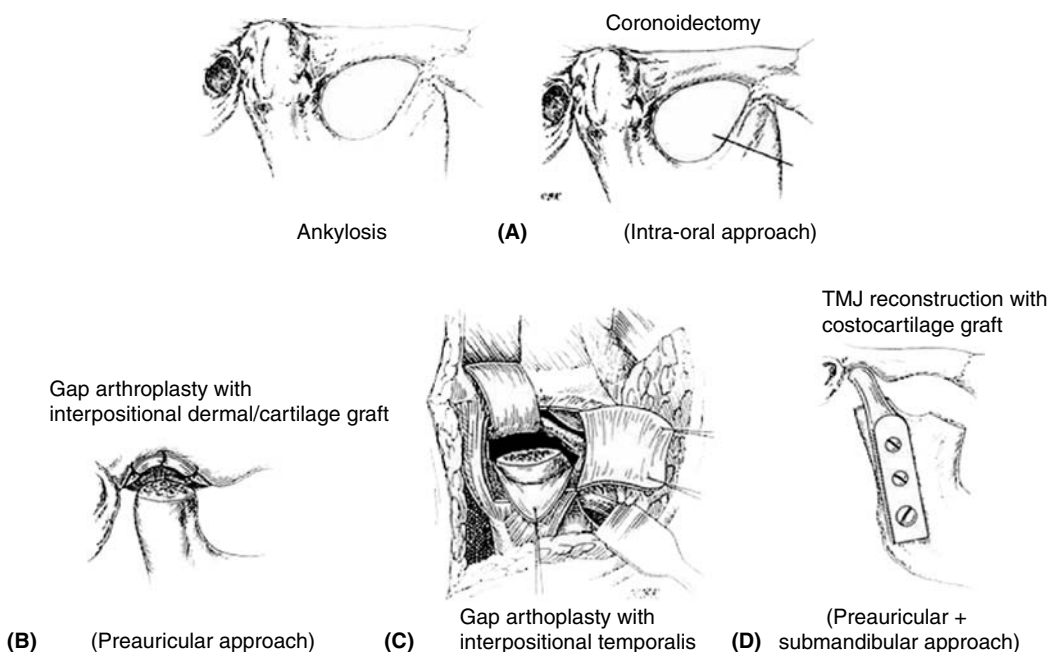


FIGURE 10 Surgical options in TMJ ankylosis: (A) coronoidectomy; gap arthroplasty (B) with dermal/cartilage graft or (C) with temporalis flap; and (D) replacement with costocartilage graft. *Abbreviation:* TMJ, temporomandibular joint.

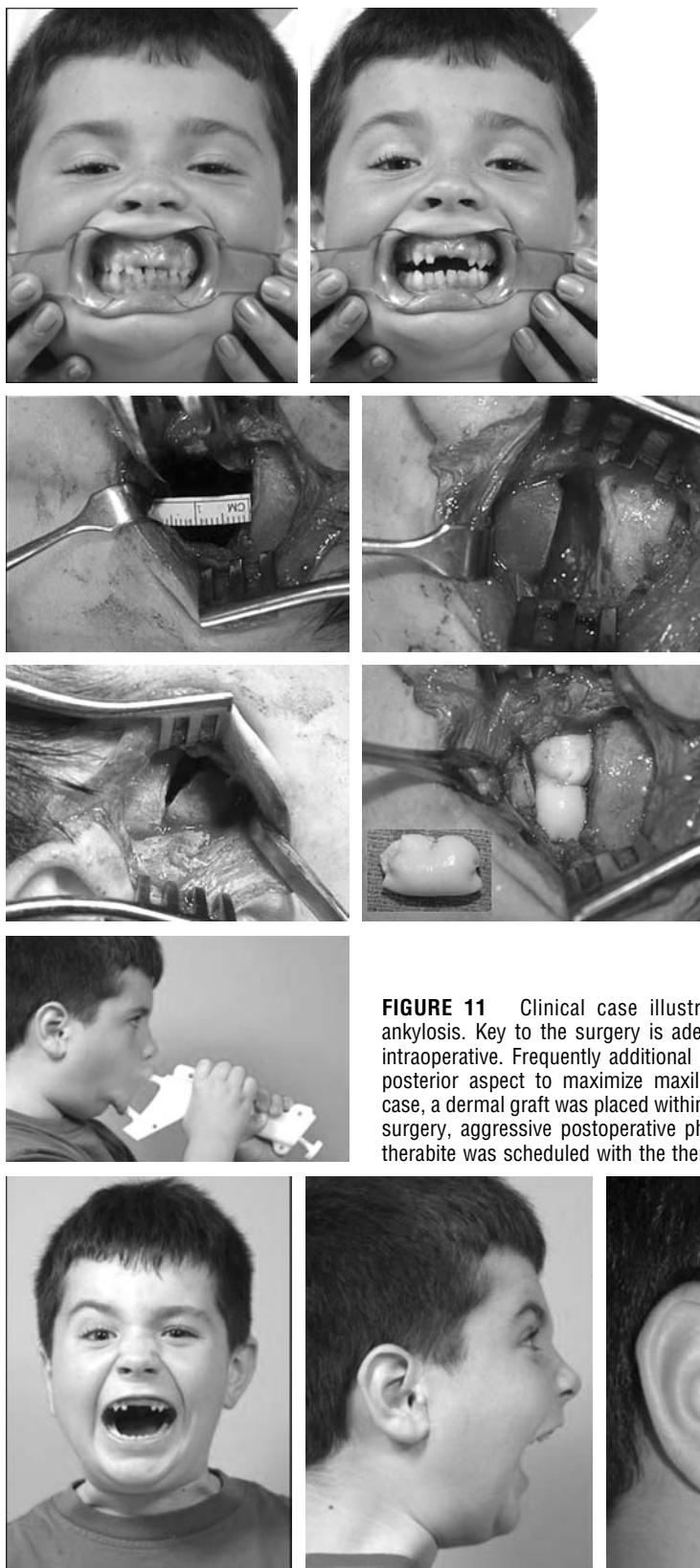


FIGURE 11 Clinical case illustrating temporomandibular joint ankylosis. Key to the surgery is adequate resection and mobilization intraoperative. Frequently additional bone must be removed from the posterior aspect to maximize maxillary-mandibular opening. In this case, a dermal graft was placed within the gap arthroplasty. Prior to the surgery, aggressive postoperative physical therapy with the use of a therabite was scheduled with the therapist.

Developmental Condylar Hyperplasia

Idiopathic condylar hyperplasia is unilateral and results in mandibular asymmetry that is asymptomatic (Fig. 12). The onset is early adolescence when it is first recognized and readily distinguishes itself from the congenital asymmetry of hemifacial microsomia and associated auricular pathology. Two distinct patterns exist. Type I or hemimandibular elongation is the most common. The condyle is of normal size and shape with overgrowth at the cartilage–bone interface resulting in an excessively long condylar neck. There is a horizontal growth resulting frequently in a midline shift with crossbite. A vertical ramal growth may coexist, but predominately the vector is horizontal. In the less common, Type II or hemimandibular hypertrophy, the condyle itself is enlarged and the mandibular ramus elongates resulting either in a maxillary-mandibular cant or an open bite. The condition is not neoplastic and is self-limiting. Diagnosis is by dentofacial films, CT, and a technetium-99m nuclear scan. In cases of active diseases confirmed by bone scan, removal of approximately 5 mm of the superior condylar surface is adequate without the need for condylectomy. The intracapsular approach should be reserved for rapid progression. However, as the disease is self limiting, the practical alternative is to allow the disease to progress to end-stage and correct using conventional orthognathic surgical techniques.

Condylar and Subcondylar Fractures

Fractures involving the condylar process of the mandible are not uncommon because of the relative structural weakness of the head and neck regions. The fractures may occur within the joint capsule or extracapsular involving the subcondylar region. The intracapsular fractures frequently involve the articular surface of the head, vascularity likely compromised resulting in avascular necrosis and the end stage is OA. Laceration, avulsion and displacement of the disc may occur. Hemoarthrosis will result in organizational scarring of the intracapsular contents with progressive or chronic joint inflammation and degeneration. Reduction of intracapsular fractures is technically difficult as fixation is impossible and the surgery itself may lead to further vascular compromise. The outcome of condylar fractures in children is more tolerant because of the remarkable ability for healing and remodeling to occur. CT followed by MRI will accurately document the pathology. Treatment is focused on regaining early ROM and occlusion to be followed expectantly. If the occlusion is altered, it typically presents with premature ipsilateral posterior molar contact and contralateral anterior open

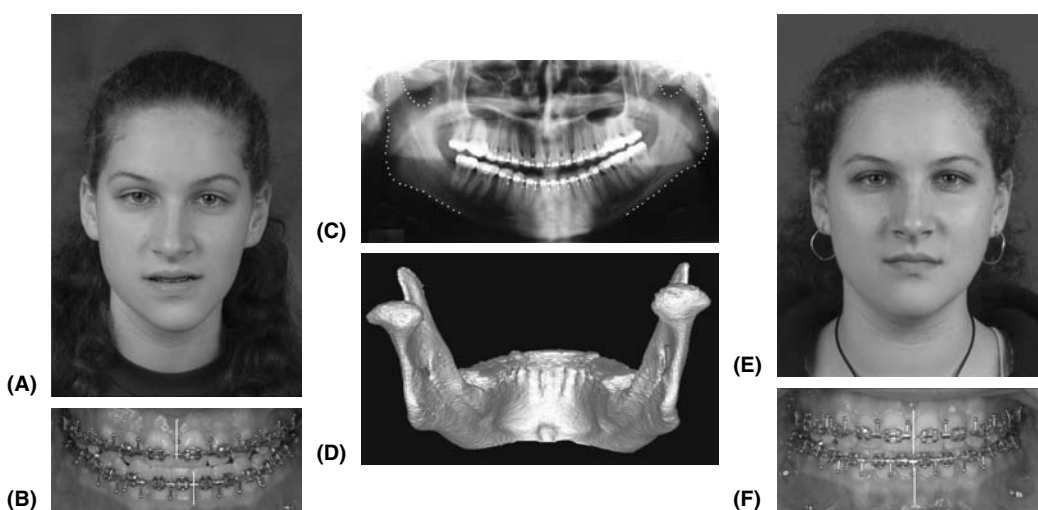


FIGURE 12 Clinical case illustrating unilateral hyperplasia. (A) Facial asymmetry, (B) occlusal maxillary-mandibular dental midline asymmetry, (C) panorex, and (D) 3-dimensional computed tomography scan showing mandibular skeletal asymmetry. Correction of asymmetry after orthognathic surgery (E) and (F).

bite as loss of the posterior height. Maxillary-mandibular fixation should be limited to two weeks, followed by guiding dental elastics, and thereafter physical therapy to regain opening and protrusion.

In extracapsular condylar neck/subcondylar fractures, the pull of the lateral pterygoid muscle will displace and angulate the proximal condyle anteromedially. In general, most fractures will do well with conservative treatment: maxillo-mandibular fixation with the addition of an occlusal splint that increases the posterior height. Treatment is typically between two and four weeks followed by guiding elastics and progressive physical therapy. However, there are indications to consider open treatment. These include when the condylar head is displaced into the medial cranial fossa, the displaced condyle blocks mandibular opening and closing, anteromedial angulation greater than 45° with dislocation of the head from the glenoid, loss of posterior vertical mandibular height with an anterior open bite deformity, and unsatisfactory outcome from conservative treatment. The decision to open is not absolute but is dependent on the circumstances and on the surgeon's experience. The surgical approach is described above, and the management is detailed in Chapter 19.

Occlusion should be followed expectantly. Secondary occlusal changes may better be addressed with orthodontic-surgical management in conventional orthognathic surgery. Whether condylectomy and replacement is needed is dependent on the goals and surgical judgment. In long standing posttraumatic dysfunction arthroscopy to assess the articular surfaces and the disc may be invaluable.

IJD Surgery

While indications for surgical management of the above structural conditions is undisputed, less clear is the role of surgery in by far the more common conditions of TMJ pain and dysfunction affecting the general population as a result of IJD and DJDs (Fig. 13). Less than 5% of all patients with TMD will eventually undergo surgery. While TMJ, MRI, and arthrography can be valuable in assessing internal derangement of the disk, regrettably the clinical symptoms do not always necessarily correlate with the pathology that is radiographically revealed. Reliance solely on such diagnostic studies without good clinical correlation and judgment may result in over diagnosis and over aggressive surgical intervention. For such conditions the indications for surgery are relative rather than absolute. The need for surgery is based on the degree of the patient's disability weighed against the outcome of nonsurgical treatment modalities described above. Thus, indications for surgical intervention are when the pain is specifically localized to the TMJ with loading and movement of the TMJ, when there is mechanical interference of the joint function, when it is refractory to nonsurgical modalities and when it results in significant impairment in the patient's daily activities.

Various surgical procedures exist for management of internal derangement. Of these the least invasive is arthrocentesis and lavage. The technique under local anesthesia involves introducing two narrow gauge needles into the superior synovial joint space through one of which Ringer's lactate is instilled with the second needle providing the outflow. Lysis of adhesions occurs under intermittent hydraulic pressure by controlling the outflow and the inlet pressure. At the end of the lavage, steroid solution is injected to minimize sequela intracapsular inflammation. This simple technique of lavaging the upper joint space alone appears to be efficacious at decreasing pain and reestablishing the normal range of mouth opening in patients with closed-lock TMJ without the need for surgically repositioning the disc. Operative arthroscopy allows a greater range of intervention, not only lavage of debris and lysis of adhesions with visual confirmation, but repositioning the displaced disk by plication and addressing surface pathology (Table 2). In contrast, open arthrotomy or TMJ surgery allows the surgeon an unlimited number of procedures to address not only intracapsular but extracapsular TMJ pathology (Table 3). The surgical approach to the capsule is described above, but the specific details of intracapsular surgery to perform the various procedures are best left to specific texts on TMJ Surgery.

Management of TMJ internal derangement

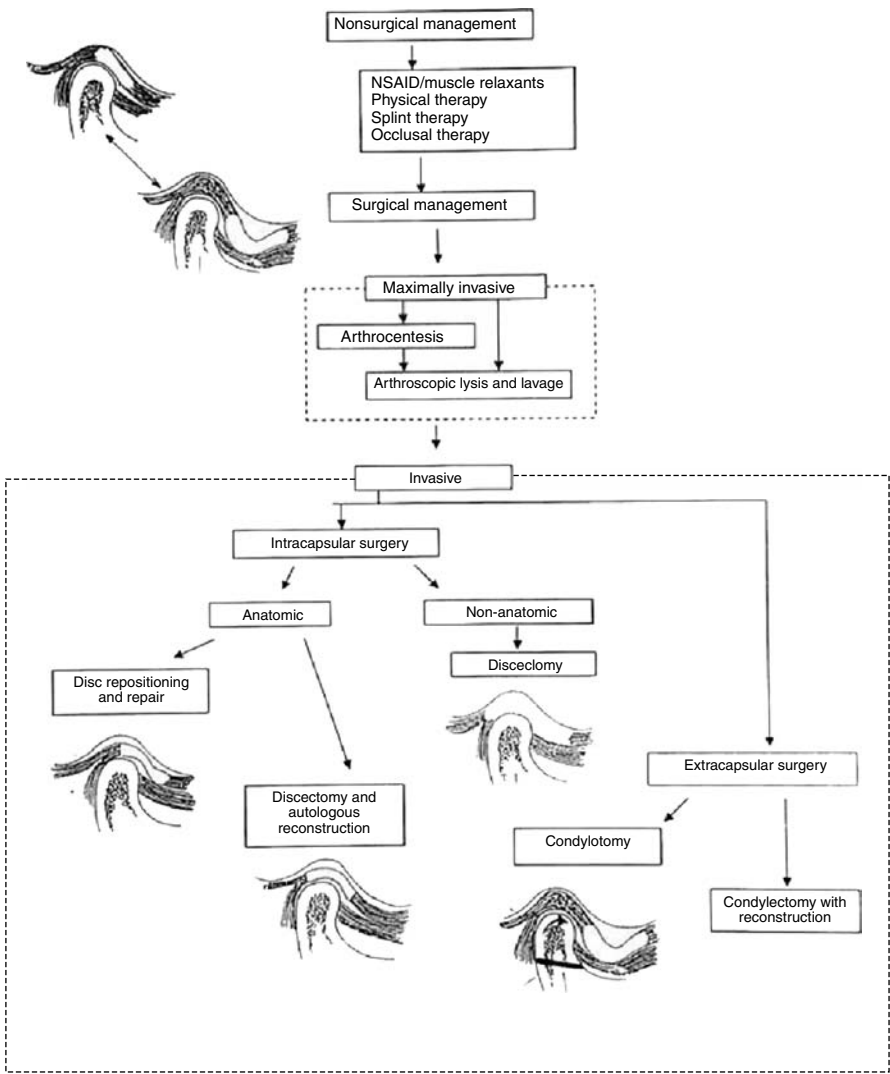


FIGURE 13 Management of internal temporomandibular joint derangement. Treatment progresses from nonsurgical modalities to minimal invasive techniques to formal surgical management of the disc and condyle. *Abbreviation:* NSAID, non-steroidal anti-inflammatory drug.

The surgical approach to the capsule is described above. Typical instruments needed for the TMJ surgery. Opening the capsule (Fig. 14) begins by instilling local anesthetic with a vasoconstrictor into the superior joint space. Key to the procedure is distracting open the joint space in a controlled manner. This is accomplished with the use of either Juniper forceps or a Wilkes spreader fixed into positioned by the use of Kirschner wires driven into the zygoma and the condylar neck. Gonial traction has been described via the

TABLE 2 Operative Arthroscopy

Lavage of the superior joint space	
1. Space	4. Contouring of surfaces
2. Lysis of adhesions	5. Disc placcation
3. Removal of debris	6. Tissue biopsy

TABLE 3 TMJ Arthrotomy

-
1. Lavage and debridement of superior joint space
 2. Glenoid fossa
 - a. Osteoplasty—lateral lip reduction
 - b. Curettage and contouring of the articular surface
 - c. Alloplastic implant
 3. Articular eminence
 - a. Eminoplasty—reduction, augmentation
 - b. Eminectomy—partial, subtotal, total
 4. Articular disc
 - a. Lysis of adhesions
 - b. Anatomic repositioning
 - c. Repair of perforation
 - i. Bilaminar flap repair
 - ii. Autogenous graft (dermal, cartilage)
 - d. Discectomy with and without autogenous cartilage replacement
 5. Condylar head
 - a. Condylotomy
 - b. Condylectomy
 - i. Without replacement
 - ii. With replacement—autogenous or alloplastic
-

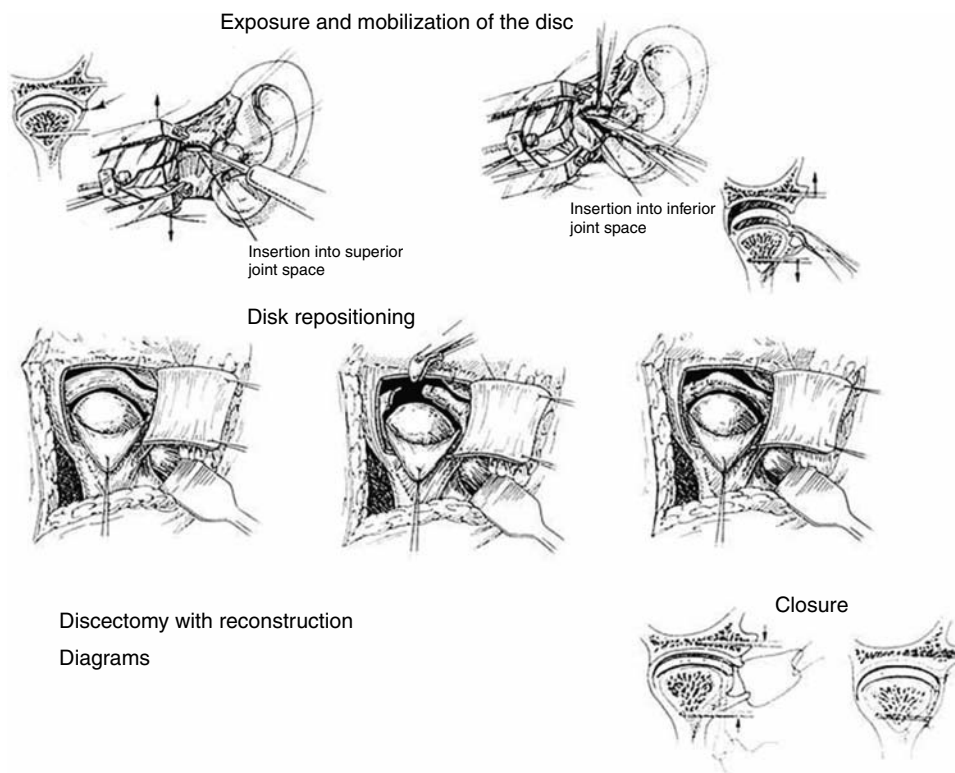


FIGURE 14 Arthrotomy: details of intracapsular surgery. The intraoperative decisions include disc repositioning, repair of perforation and replacement or its removal. Additionally, the surface of the condyle can be contoured depending on the pathology with this exposure.

submandibular approach to distract the joint, but this is not practical because of the mobility of the head and the inability to control the spatial gap in a controlled manner. The spreaders are critical and must be well secured before proceeding. The inferior border of the lateral lip of the glenoid fossa is palpated and a curved incision made through the lateral capsule leaving a cuff of 2 to 3 mm to allow repair of the capsule at the time of closure. With fine scissors the incision is extended along its length and then directed superiorly into the upper joint space to minimize injury to the disk. As the exposure is made, the spreader forceps is used to further distract the joint cavity for visualization. The characteristic shape of the disc is visualized between the posterior and anterior bands. To enter the lower joint space, the disc is grasped with a forceps and retracted superiorly to place the lateral collateral ligament in tension. The lateral ligament is then divided and the lower joint space entered. The spreader is further distracted open and intracapsular elements can now be visualized to be addressed surgically. The integrity of the disc and adhesions is accessed. Anterior and medial adhesions in the upper joint space usually restrict the mobility of the disc in long standing disc displacement. With the disc retracted posterolaterally, these must be carefully released without perforating the medial capsule by sharp dissection and the use of a curved freer until the disc can be mobilized. Grasping the disc to provide counter traction must be done with care without crushing the disc itself. Instead the disc should be grasped with fine forceps where the tissue that will be resected or pliated. With the disc repositioned near its anatomic position over the condyle, the redundant tissue of the posterior band can be resected and reapproximated. The spreader should be closed down when determining the length of redundancy. Next the lateral fixation of the disk is accomplished by a series of horizontal mattresses so that disc is pulled into the sulcus between the condyle and the lateral capsule. In situations where there is disc perforation, this is addressed by resecting the margins of the perforation, mobilizing posterior laminar regions separately between the strata and approximating the edges. An alternative is to "patch" the perforation with a dermal graft. When an adequate repair cannot be performed, either discectomy with or without autogenous auricular cartilage graft is an option. Regardless of the specifics of the intracapsular surgery, postoperative management with modification of the preoperative splint therapy and physical therapy to regain ROM is essential and cannot be over emphasized.

An alternative surgical approach to the management is a modified condylotomy. The approach is intraoral and is described as above. The concept here is instead of repositioning the disk, the osteotomized condyle itself is repositioned beneath the anteriorly displaced disc. The joint space is increased and effectively the condyle–disc–fossa relation is reestablished. Postoperatively the patient is maintained in prolonged maxillary-mandibular fixation followed by training elastics and splint therapy.

CONCLUSION

While the outcome may be variable, the surgical decision making algorithm where the TMJ is affected as a result of congenital deformities or acquired as in ankylosis, fractures and tumors is relatively straight forward based on morphologic restoration of the "gross" anatomy. The basic tools are within the armamentarium of craniofacial surgeons. However, TMJ surgery that focuses on intracapsular pathology is in itself a subspecialty, requiring the surgeon to make a commitment to become not only technically proficient in the various surgical options of TMJD but also gain an understanding of the nonsurgical modalities that are the main stay of treatment for the vast majority of patients. Ultimately the decision to surgically intervene is based on the surgeon's judgment and experience in his/her ability to provide relief for the patient with what is available in his/her armamentarium. The predisposing, inciting and perpetuating factors of TMJD are varied. As the causes of TMJ disorders are multifactorial, the surgeon is only one member of a team consisting of general dentists, orthodontists, physical therapists and psychologists. Even if surgery is indicated, failure to include nonsurgical management (medical, occlusal,

physical therapy and psychological support) to support surgical intervention before and after surgery is to invite clinical failure though surgery itself may be technically flawless.

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16 Craniofacial Microsomia

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INTRODUCTION

Craniofacial microsomia, or otomandibular dyostosis, is a spectrum of soft and hard tissue hypoplasia typically affecting the regions of Tessier's #6, #7, and #8 facial clefts (1). It is a disorder of the first and second branchial arches with an incidence as high as 1 in 3500 live births. The classic presentation is unilateral mandibular hypoplasia, deficient soft tissues of the face and microtia. Gorlin's term, hemifacial microsomia, emphasizes that the disorder is usually a unilateral facial deformity (70–95% of the time) and should not be confused with Treacher Collins Syndrome (also a Tessier's #6, #7, and #8 facial clefts), which is bilateral and symmetric (2).

VARIATION

Craniofacial microsomia is known for its wide spectrum and variation that ranges from the mild forms, with only preauricular skin tags, to moderate forms, which may have a hypoplastic mandible and diminished cheek fat but a near normal external ear, to the severe form of Goldenhar Syndrome (oculovertebral sequence). Goldenhar syndrome is a severe variant of craniofacial microsomia defined by colobomas of the eyelid and cervical vertebral anomalies, as well as the other head and neck sequelae of craniofacial microsomia, including mandibular hypoplasia, macrostomia, and microtia.

ASSOCIATED ANOMALIES

The primary anomalies of hemifacial microsomia are that of mandibular hypoplasia, microtia, and deficient soft tissues of the face. These patients also have a high incidence of preauricular skin tags, conductive hearing loss, hypoplastic glenoid fossa, and cranial nerve VII palsy. Also associated are macrostomia and masticatory muscle hypoplasia. Some of these patients are born with upper airway obstruction related to micrognathia, with related posterior tongue-base collapse. Significant upper-airway obstruction may require tracheostomy or early mandibular intervention to improve the mandibular position. Many of the patients with craniofacial microsomia and upper airway obstruction also have symptomatic gastroesophageal reflux. There is a high incidence of bony midfacial deformities mostly seen as maxillary and zygomatic hypoplasia contributing to these patients' occlusal cants. Temporal and frontal deficiencies may also be seen. Cleft lip and/or palate have been reported.

CLASSIFICATION

Encompassing the orbit, mandible, ears, nerves, and soft tissue, the OMENS classification was developed as a global classification (3). While the OMENS classification addresses the whole face, the Pruzansky classification is used for the abnormal mandible, and the Muerman classification is for the external ear pathology.

Mandibular pathology is the most evident and occurs to some degree in 89% to 100% of the patients with craniofacial microsomia. The Pruzansky classification is commonly used to

document mandibular deficiency (4). There are four types relating to severity of the mandibular deformity:

Pruzansky Classification

- *Type I* Mild ramal shortening.
- *Type IIA* The condyle and ramus are small, but the condyle and glenoid fossa are anatomically oriented. The coronoid may be absent.
- *Type IIB* The condyle and ramus are small and medially displaced, and there is no functional glenoid fossa. (The subdivisions IIA and IIB are based on the TMJ pathology.)
- *Type III* Complete absence of the ramus, condyle, and coronoid process.

MANDIBULAR SURGERY

The mandible can be operated at all stages of the child's life depending on necessity of the operation. If airway obstruction exists during the newborn period, or manifests later as obstructive sleep apnea, then mandibular distraction may be performed early. In addition, mandibular distraction may be performed in young children to facilitate decanulation for a tracheostomy. If there are no functional issues, and there is significant mandibular ramal asymmetry, mandibular distraction is usually performed between the ages of six years to eight years.

Pruzansky type I mandibles may not require distraction osteogenesis during mid-childhood. In these patients, orthodontic treatment with functional appliances may be helpful during growth. Then, at skeletal maturity, an orthognathic procedure may be required. For Pruzansky type IIA and IIB, distraction is typically helpful during mid-childhood, with slight over-correction being the goal. Even with this over correction, an orthognathic procedure will be necessary in adolescence.

Operative Technique: Mandibular Distraction

For the operative technique, although an external device may be used, an internal distraction device is preferred for decreased scarring and patient comfort (see Chapter 14). An intraoral exposure and oblique osteotomy is used, as opposed to the neonatal procedure that uses a risdan incision and inverted "L" osteotomy.

For Pruzansky type III mandibles, a costocartilage or rib graft may be used (Fig. 1). Alternately, distraction may also be used in these patients; however, a three dimensional computed tomography (CT) scan should document a bone segment sufficient to anchor the cephalad screws or pins. Some clinicians have staged this procedure with molar tooth bud extraction followed by distraction of a Pruzansky type III mandible (5). Sacrificing teeth for this reason when a rib graft is a viable option is controversial.

Survival and growth of a rib graft is unpredictable. At times, a rib graft even overgrows, requiring a set-back at skeletal maturity. Other times the rib graft may dissolve and require regrafting. Once healed, rib grafts may be lengthened with distraction. The osteotomy site for distraction of a rib graft should be either on the native mandible or on the rib graft. Complications and even loss of the rib graft may result if the healed juncture between the rib graft and native mandible is used as the osteotomy site.

Operative Technique: Costocartilage Graft for Mandible Reconstruction

1. The rib graft is harvested through an oblique 4 cm anteriolateral chest incision.
2. The 8th, 9th, or 10th rib is exposed, and a Doyen rib dissector is used to strip the soft tissues.
3. Anteriorly, a 1 cm cartilaginous cap is harvested, along with 10 cm to 12 cm of bone rib graft, with a knife anteriorly and a bone cutter posteriorly.
4. Valsalva maneuver is used to check for pleural injury prior to a layered closure.
5. The cartilage cap may be secured with a 90-90 2-0 PDS suture. Small corticotomies may be made with a diamond burr throughout the rib graft to allow for vascular ingrowth.

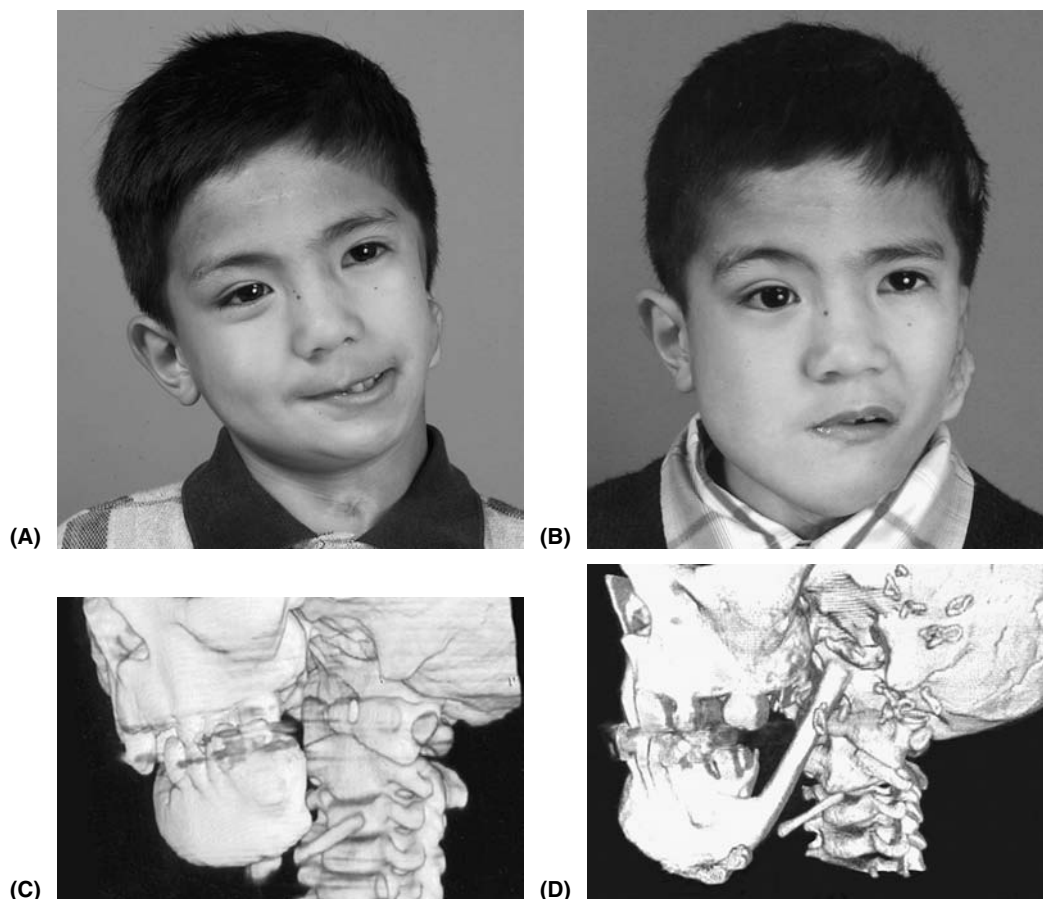


FIGURE 1 Images of a patient with left Goldenhar syndrome. (A) Preoperative image demonstrates severe left mandibular hypoplasia. (B) Postoperative image following a costocartilaginous rib graft shows improved chinpoint position. (C) Preoperative computed tomography (CT) scan demonstrates a Pruzansky type III mandible with no ramus or temporomandibular joint. (D) Postoperative CT scan shows consolidated rib graft.

6. Through a 3 cm submental incision, subperiosteal dissection is performed to expose the mental and parasymphyseal region of the affected side.
7. A tunnel is made with a tonsil clamp toward the glenoid fossa or cranial base. The tunnel is widened to accommodate the surgeon's index finger.
8. The rib graft is directed through the enlarged tunnel up to the cranial base.
9. A large bone hook is used to pull the mandible to the contralateral side during fixation.
10. Lag screws (two 12–15 mm by 2 mm) are used to fix the rib graft to the inferior portion of the mandible and the submental incision is closed in layers.

Orthodontic Therapy

Orthodontic therapy is important during the treatment period in mid-childhood for distraction of the mandible and during adolescence for orthognathic preparation. At the time of mandibular distraction, guiding elastics are often used during the consolidation phase of distraction to mold or shape the generate bone and close an open bite. In preparation for orthognathic surgery the orthodontist may have to upright abnormally inclined teeth, establish an appropriate maxillomandibular arch-width relationship, and level the occlusal plane.

MAXILLARY SURGERY

As previously mentioned with unilateral involvement, the maxilla is short on the affected side and long on the contralateral side. In mid-childhood, mandibular distraction is aimed at correction of the deficient mandibular ramus, not the maxillary asymmetry. However, the lateral open bite that develops on the affected side post-distraction generally closes over a six to twelve week period with extrusion of the maxillary teeth. Another procedure that has been described is distraction of the coupled maxillomandibular units (after Le Fort I and bilateral sagittal split osteotomy) during mixed dentition (6). However, we have not found this bimaxillary distraction procedure necessary.

In a skeletally mature patient, a double jaw procedure or Le Fort I osteotomy with simultaneous bilateral sagittal split osteotomy of the mandible is used to correct an occlusal cant and move the chinpoint to midline (Fig. 2). In addition, osseous genioplasty is useful at the time of this procedure to correct asymmetries because it allows for movement of the chin-bone segment in 3 dimensions or 6 directions (anterior-posterior, vertical, horizontal).

In preparation for orthognathic correction, radiographic studies and dental models are obtained and intermediate and final splints are fashioned (see Chapter 13). The malposition (or absence) of the external auditory canal in some patients with craniofacial microsomia is unique in obtaining preoperative records. Careful placement of the facebow registry without ear tongs in the malpositioned or absent external canal is, thus, necessary.

FRONTO-ORBITAL-ZYGOMATIC ANOMALIES

Only the more severe patients benefit from surgical correction of asymmetries after lower facial corrective surgery has been performed. Vertical orbital dystopia may be addressed with orbital box osteotomy or facial bipartition (Fig. 2).

EAR CLASSIFICATION

With craniofacial microsomia, 66% to 99% of the patients have microtia. Other abnormalities of the ear include conductive hearing loss, middle ear deficiencies, and preauricular skin tags. There are three grades of auricular anomalies based on Meurman classification:

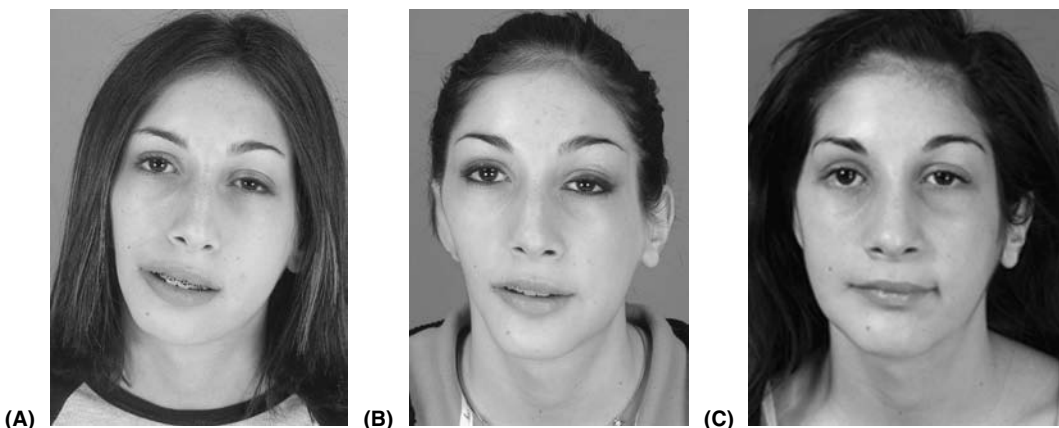


FIGURE 2 Frontal images of patient with right craniofacial microsomia. **(A)** Preoperatively, her occlusal cant and significant facial asymmetry is noted. **(B)** After orthognathic correction from Le Fort I osteotomy and bilateral sagittal split osteotomy, the chinpoint is corrected to midline, but right soft-tissue hypoplasia persists. **(C)** After autologous fat grafting to right cheek and inferior translocation of right orbit using an orbital box osteotomy. Improved symmetry is achieved.

Meurman Classification

- *Grade I* Normal shape; all components of the ear are present but deficient.
- *Grade II* There is only a vertical remnant of cartilage and skin.
- *Grade III* Only a lobule is present; some patients have anotia.

TOTAL EAR RECONSTRUCTION

First stage total ear reconstruction is begun between five and eight years of age. We use either the 2-stage Firmin modification of the Nagata procedure or the 4-stage Brent technique, depending of the severity, lobule location, and skin availability (7,8). For a majority of the cases of microtia, the Firmin technique of framework fabrication and ear elevation is used (Fig. 3). This framework includes four pieces of rib cartilage (the base, helix, antihelical fold, and tragus) secured together. Compared to the Brent framework, the Firmin framework is larger and at slightly higher risk for exposure. However, this technique potentially gives a superior result. Six months after framework placement, ear graft elevation is performed, with banked cartilage graft covered with a temporoparietal fascial flap and full-thickness skin graft.

The Brent technique is used with a low, malpositioned ear lobule or shortage of skin (Fig. 4). Proper locations of the external ear reconstruction, related to the contralateral ear, is even more important than a detailed framework. The Brent technique is staged: (i) cartilage framework placement, (ii) lobule rotation, (iii) framework elevation with banked cartilage and FTSG, and (iv) tragus reconstruction from a contralateral conchal bowl skin/cartilage composite graft.

After final completion of the total ear reconstruction, the external auditory canal reconstruction may be performed. A CT scan with fine cuts through the mastoid region helps the head and neck surgeon identify the location of the facial nerve to avoid injury. The canal is formed by drilling out a passage through the mastoid bone and lining it with a skin graft. For bilateral microtia, a removable hearing aid is used until four or five years of age, when an osseointegrated bone-anchored hearing apparatus (BAHA) may be placed.



FIGURE 3 Total auricular reconstruction with two-stage Firmin technique (inset: close-up). (A) Preoperative; (B) postoperative after first-stage four-piece rib-cartilage framework fabrication.



FIGURE 4 Total auricular reconstruction with 4-stage Brent technique. (A) Preoperative; (B) postoperative after rib-cartilage framework fabrication, subsequent lobule rotation, and then tragus reconstruction.

SOFT TISSUE

With craniofacial microsomia, the soft tissue cover of the involved region is deficient, including: the skin, muscle, and fat layers. Soft tissue augmentation for facial symmetry in unilateral craniofacial microsomia is as important as the hard tissue or bony reconstruction. We prefer to use serial fat transplantation performed each time a patient has anesthesia for staged reconstruction. Since many patients with moderate or severe involvement require multiple operations throughout their childhood for mandibular distraction, staged ear reconstruction, orthognathic procedure, and others, there are many opportunities for serial fat transfer. We use the Coleman technique of nontraumatic, blunt-tipped fat harvest and small aliquot, layered fat injection. If a large volume of soft tissue augmentation is required at skeletal maturity, then we use the inframammary extended-circumflex scapular (IMECS) or parascapular fascial fat-free flap (8). This flap is effective, but requires revisionary procedures to gain symmetry.

FACIAL PALSY

Deficiency of the facial nerve (VII) can be seen. Most common is a non-functioning marginal mandibular branch. The degree of facial nerve weakness does not necessarily correlate with the degree of hard or soft tissue deficiency. Mild to moderate weakness, not complete facial nerve paralysis, is typically observed. Thus, a cross-facial nerve graft and staged gracilis muscle-free flap or other reconstructive facial reanimation surgery is rarely needed.

TIMING OF TREATMENT

The treatment protocol for a patient with craniofacial microsomia may be individualized depending on the regions of involvement. The timing of treatment is dependent on functional needs, age, growth, and development (Table 1).

TABLE 1 Craniofacial Microsoma: Timing of Treatment

Age	Indications	Operative procedure
Birth–18 months	Upper airway obstruction (significant)	Tracheostomy or neonatal mandible distraction
	Macrostomia	Macrostomia correction: modified Z-plasty w/orbicularis oris repair
	Other anomalies: cleft lip, cleft palate, preauricular skin tag	Correction of other anomalies
18 months–3 years	Tracheostomy	Mandibular DOG for decannulation (Pruzansky IIA or IIB)
4–13 years	Mandibular asymmetry	Mandibular DOG (Pruzansky IIA–IIB) Rib graft (Pruzansky III) (5–8 years)
	Microtia	Staged total ear reconstruction (5–8 years)
Adolescent–adult	Malocclusion (occlusal cant)	Double jaw: Le Fort I and BSSO of mandible
	Soft-tissue hypoplasia	Serial fat grafting IMECS free flap

Abbreviations: BSSO, bilateral mandibular ramus sagittal split osteotomies; DOG, distraction osteogenesis; IMECS, inframammary extended-circumflex scapular.

SUMMARY

Craniofacial microsomia is not an uncommon deformity, with variable facial involvement and functional disturbances, including respiratory and hearing difficulties. Hard- and soft-tissue correction involves multidisciplinary planning of the proper timing and operative technique.

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17 Treacher–Collins Syndrome

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INTRODUCTION

Treacher–Collins syndrome was originally described in 1846 by British ophthalmologist Edward Treacher Collins. Franceschetti et al. coined the term mandibulofacial dysostosis in 1944. By 1969, Tessier described this syndrome as a bilateral confluent cleft numbered 6, 7, and 8 using his craniofacial cleft classification system (1).

The fully expressed Treacher–Collins phenotype has deficiencies and malformations of both the craniofacial skeleton and the surrounding soft tissues of the face, the external ear, and hearing apparatus (Fig. 1) (2–4). Features may include hypoplasia or aplasia of the zygomatic body and arch, mandibular ramal hypoplasia, increased facial convexity, and a retrusive chin with increased vertical height. The disorder is transmitted in an autosomal dominant fashion with complete penetrance and variable expression. The incidence ranges from 1 in 10,000 to 1 in 50,000 live births. The site of transmission in some cases has been mapped to chromosome 5q31.3 to 33.3, whereas other cases appear to have a deletion in chromosome 4p (5–7). Up to 60% of cases may occur without a family history and, therefore, represent spontaneous index cases (8).

FEATURES

The obligatory features described by Munro include midfacial deformities: (i) downward slanting palpebral fissures, (ii) coloboma or cleft of the lower lateral eyelid including eyelash abnormalities, and (iii) malar hypoplasia (Table 1). Mandibular defects, micrognathia, and fish-like facial appearance complete the obligatory features of Treacher–Collins syndrome (Table 2) (9). Associated but nondefining features include macrostomia, anteriorly displaced preauricular hairline, malocclusion with open bite, cleft or arched palate, and auricular malformations. Distinguishing element to Treacher–Collins syndrome is that it is *bilateral and symmetrical*; however, slight asymmetry may be present (10,11).

The absent or diminutive zygoma represents the major feature of this syndrome. A rudimentary zygomatic arch without connection to the maxilla or frontal bone is often the only malar structure (4). An oval-shaped orbit with a large inferior and lateral cleft with communication to the inferior orbital fissure and associated herniated soft tissue accompanies the absent zygoma (2,4). Evaluation of the maxilla reveals a narrow and shortened posterior aspect, a high palate with or without a cleft, and an overprojected anterior portion (3). The maxillary features further accentuate the deficiencies in the zygomatic region. Class II malocclusion with an associated open bite is often seen. An obtuse gonial angle, a short vertical ramus, and a concave antgonial notch are unique features of Treacher–Collins syndrome. Choanal atresia and pharyngeal hypoplasia along with the diminutive mandible are associated with airway obstruction in this syndrome (4).

The lower eyelid cleft or coloboma is thought to be indicative of Treacher–Collins syndrome (4). Often the cilia are absent from the lower lid or its medial two-thirds. The loss of orbital domain, lack of lateral orbital wall, and the herniated orbital contents result in the

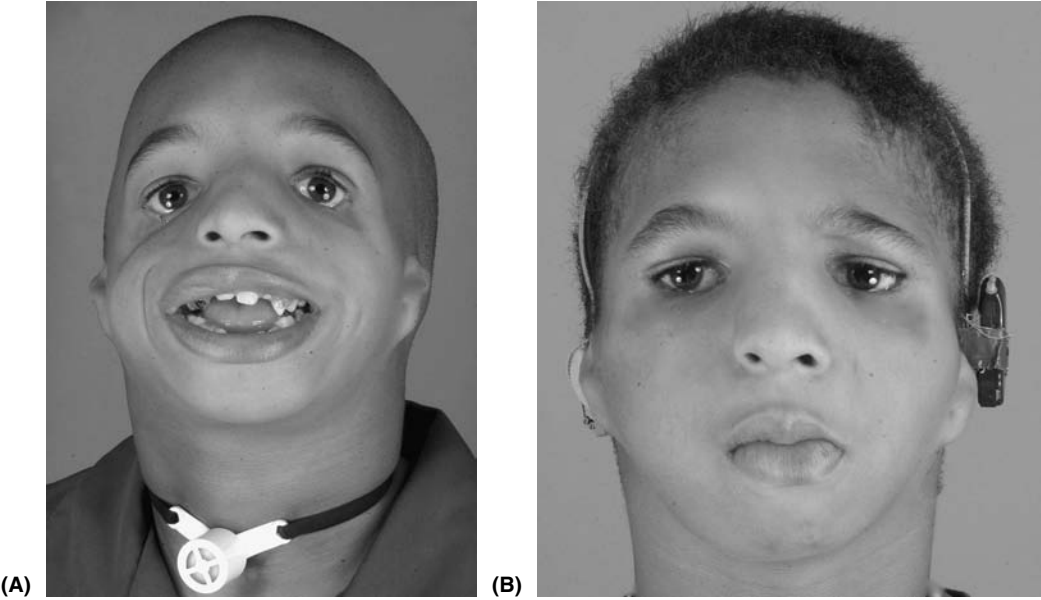


FIGURE 1 A nine-year-old Treacher–Collins syndrome patient, who is tracheostomy dependent, has micrognathia, bilateral microtia, zygomatic hypoplasia, and antimongoloid slant of lateral orbital eyelid region. **(A)** Frontal preoperative view. **(B)** Frontal postoperative view after mandibular distraction, removal of tracheostomy malar reconstruction with cranial bone grafts, and eyelid switch flaps.

downward lateral slant of the palpebral fissure (2). The sideburns are often long, anteriorly displaced, and extend inferiorly in a tongue-like fashion. A cheek furrow or occasionally a full-thickness defect extending from the commissure to the lateral orbit can be present.

The ear deformities are usually bilateral and symmetric. Microtia rates of 85%, external auditory canal stenosis of 54%, and auditory canal atresia as high as 31% (3,10). The middle ear is also abnormal with hypoplastic cavities present and small, fused, or absent ossicles. The majority of patients with Treacher–Collins syndrome have some degree of hearing loss mainly of the conductive type. The incidence of hearing loss has been reported as high as 96% (12).

Although patients presenting with full expression of Treacher–Collins syndrome are readily diagnosed, mild expression may prove a diagnostic challenge. Bilateral craniofacial syndromes that may confused Treacher–Collins syndrome include bilateral hemifacial microsomia, Goldenhar syndrome, Nager or Miller syndrome, and Hallerman–Streiff syndrome (13–15). Distinguishing features such as ocular and lower eyelid coloboma and loss of lower lid eyelash hair are not present in hemifacial microsomia or Goldenhar syndrome. Nager syndrome and Miller syndrome are autosomal recessive syndromes with either preaxial or postaxial limb deficiencies, respectively. Patients with Hallermann–Streif syndrome are distinguished by their paucity of facial hair in the upper third of the face and by the light color of that hair.

TABLE 1 Treacher–Collins Syndrome: Soft Tissue Features

Palpebral
Antimongoloid slant of fissure
Lower eyelid coloboma
Absent eyelash on medial two-thirds or lower lid
Ear
Microtia
Conductive hearing loss
Anterior displacement of preauricular hair
Malar
Soft tissue hypoplasia (fat/muscle)

TABLE 2 Treacher–Collins Syndrome: Skeletal Features

Mandible
Retruded
Short ramus
Anterior open bite
Deep antegonial notch
Retromicrogenia
Maxilla
Reduced posterior facial height
Small posterior nasal cloana

TREATMENT

Reconstructive procedures for the Treacher–Collins patient occur during specific times of facial growth and depend on the severity and regions involved. Key operations focus on the malar region during mid-childhood with zygomatic, orbital and eyelid reconstruction, and orthognathic correction during adolescence (Table 3). However, other ancillary procedures are often required at specific timepoints as well (Table 4).

Neonatal Period

In the neonatal period treatment options for the Treacher–Collins patient begins with evaluation of the airway and the propensity for airway obstruction due to the micrognathic mandible present in more expressed phenotypes (16,17). Feeding difficulties may exist and be related to jaw abnormalities. For severe upper airway obstruction in the newborn, tracheostomy is the gold standard. However, neonatal mandibular distraction has been successfully used to avoid a tracheostomy and should be considered with a multidisciplinary team (see Chapter 11) (18,19). If other airway obstructive lesions exist, like cloanal atresia (in addition to the posterior tongue collapse from micrognathia), then a tracheotomy should be placed and neonatal distraction should not be attempted. If a tracheostomy is placed for upper airway obstruction due to micrognathia in Treacher–Collins patients, then an elective mandibular distraction procedure may be planned in order to work toward decanulation (16,20,21). This process may be difficult and additional head and neck procedures, like laser treatment of granulation tissue and tracheal reconstruction, may be necessary before successful decanulation is possible.

After airway stabilization and adequate nutrition has been achieved, early interventions including repair of associated cleft lip by three months of age and repair of cleft palate by one year may be performed. Other procedures that may be required prior to one year of age include the correction of macrostomia or exclusion of preauricular pits or skin tags.

TABLE 3 Treacher–Collins Syndrome: Treatment Protocol

Age 6–8 yr
Malar reconstruction
Cranial bone graft to zygomatic arch/orbital floor
Upper to lower eyelid switch flap/lateral canthopexy
Malar left/fat grafting
Mandibular distraction
External ear (microtia) reconstruction
Late adolescence
Orthognathic surgery
Le Fort I (posterior impaction)
Bilateral sagittal split osteotomy
Osseous genioplasty
Soft tissue augmentation

TABLE 4 Treacher–Collins Syndrome:
Ancillary Procedures

Newborn
Tracheostomy
Neonatal mandibular distraction
3 mo
Cleft lip repair
Macrostomia correction
1 yr
Cleft palate repair and myringotomy tubes
Preauricular skin tag removal
2–6 yr
Bone-anchored hearing apparatus placement

Ancillary procedures are only performed if needed.

Optimizing hearing is an important goal during this period of language acquisition. Bone conductive hearing aids are often required. In addition, a bone-anchored hearing apparatus (BAHA) may be beneficial in some cases. (If a pediatric otologist places osseous integrated screws for the BAHA, then location should be confirmed by the craniofacial surgeon so that future total ear reconstruction is not compromised.)

Although the eyelid and orbital deformity may be the most striking feature even at birth, typically correction should be delayed until mid-childhood. One important reason for this delay is to minimize the number of surgeries necessary for correction. Bone grafts used to augment the malar region placed early will dissolve. Enlow documented that this malar region is an area of bone resorption (as opposed to apposition) (22). Bone grafts to the malar region performed at a very young age will dissolve and need to be regrafted (23,24).

Mid-Childhood

In mid-childhood, during six to eight years of age, multiple procedures may be required depending on the regions and severity of the Treacher–Collins syndrome involvement. During this age range correction includes (i) mandibular distraction lengthening; (ii) malar, zygomatic, and eyelid reconstruction; and (iii) total external ear reconstruction. Both the distraction and total ear reconstruction procedures are staged and required multiple surgeries. During each of these staged procedures autologous fat grafting may be added for soft tissue augmentation. (One advantage of this incremental soft tissue augmentation throughout childhood procedures is the avoidance of a major free flap operation to provide soft tissue fill in adolescence.)

Mandibular Distraction

Severe mandibular deformity without airway compromise can be corrected during mid-childhood using distraction osteogenesis (25,26). A vertical vector should be used in placement of the distraction device to correct the short ramus and improve the obtuse gonial angle. Anterior elastic bands should be used on orthodontic hooks or maxillomandibular fixation screws to guide the generate bone and close the open bite. Relapse of this abnormal mandibular morphologic shape from the strong genioglossal muscle pull has been noted in long-term follow-up, but this may be addressed later during an orthognathic procedure.

Total Auricular Reconstruction

There are two types of total auricular reconstruction techniques that are commonly used (see Chapter 16). Brent and Tanzer described and popularized a four-stage method (27,28):

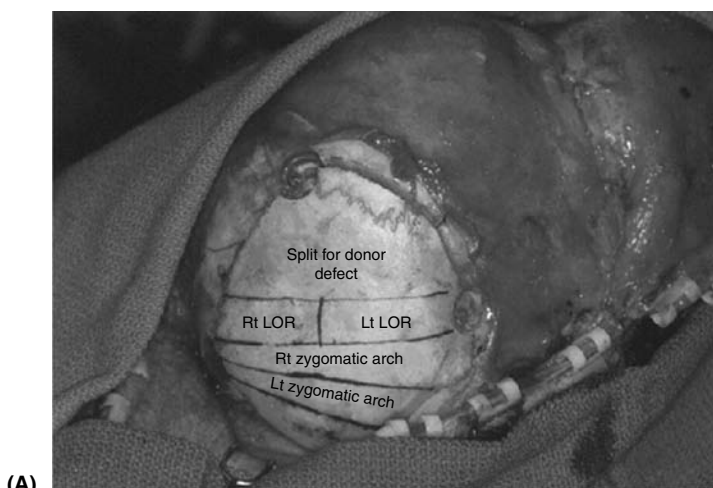
1. Rib cartilage helical rim and antehelix fabrication and implantation.
2. Elevation with banked cartilage and full-thickness skin graft.
3. Lobule transposition.
4. Tragus reconstruction with a contralateral composite skin/cartilage graft and conchal bowl reconstruction with full-thickness skin graft.

More recently, Nagata and Firmin described a two-stage total auricular reconstruction (29,30):

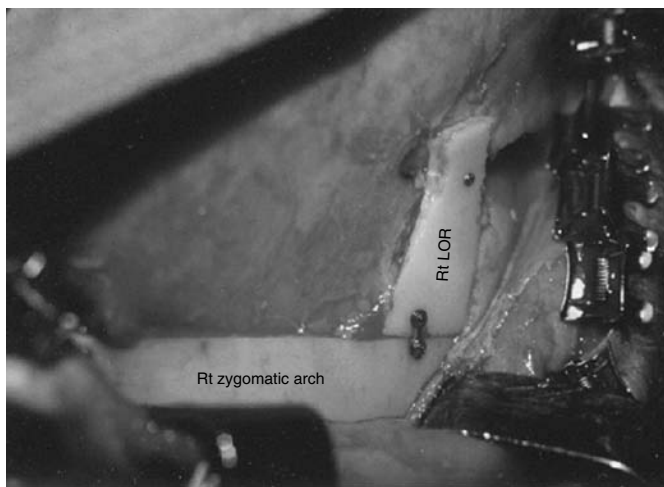
1. Rib cartilage total ear fabrication by joining 4 pieces of cartilage (the base, helical rim, antehelix, and tragus).
2. Elevation with banked cartilage graft and temporoparietal pedicle flap coverage with full-thickness skin graft.

Malar Eyelid Reconstruction

The zygomatic deficiency or central event of Treacher–Collins syndrome is also addressed during mid-childhood ages from six to eight years. Waitzman and Posnick reported near complete growth of the zygomatic maxillary orbital complex by the age of five to seven years, and therefore recommend orbital reconstruction after that age (31). Tessier also recommends orbital reconstruction after the age of six years due to resorption of bone grafts placed in infancy for the treatment of this particular syndrome.



(A)



(B)

FIGURE 2 (A) Intraoperative view of parietal skull with the vertex of the skull on the left of the image, the patient's nose positioned on top of the image, the right ear to the right of the image, and the occiput at the bottom of the image. The markings are shown for a full-thickness cranial bone graft to be divided into the right and left zygomatic arch and right and left lateral orbital rim (LOR). The remaining half is split to fill the cranial donor defect. (B) Interoperative view of reconstructed right zygomatic arch and right LOR.

Zygoma and orbital bone reconstruction using cranial bone grafts is the most common form of reconstruction (32,33). Although vascularized bone grafts have been described, there is some evidence that in an area of bone resorption, like the malar region, that resorption is actually hastened with such vascularized grafts (34). Thus, we prefer full-thickness parietal bone grafts for the zygomatic arch and lateral orbit, split thickness grafts for the orbital floor, and a simultaneous upper to lower eyelid switch flaps with lateral canthopexies, as described below.

Operative Technique: Malar/Lower Eyelid Reconstruction

1. A zigzag coronal incision is used for the exposure of the nondominant parietal skull and orbits.
2. Temporal muscle is left in the place by dissecting on the superficial layer of the deep temporal fascia to avoid the frontal branch of the facial nerve.
3. A circular craniotomy is performed in the parietal skull measuring approximately 6 cm².
4. The bone graft is divided into two (Fig. 2A). One of the halves is split through the diploic space and used to cover the cranial defect. The other half is divided into three long strips. Two of the strips are used for the zygomatic arch. *Note:* The curved portion of the parietal bone should be used for the curved portion of the anterior malar region. The remaining strip of the bone is divided in the center to make both lateral orbital walls.
5. Rigid fixation is used to secure the zygomatic arch and lateral orbit bone grafts (Fig. 2B). (A lag screw may be used at the root of the arch to secure the graft.)
6. Split calvarial bone grafts are next harvested and placed in the lateral orbital floor.
7. The lower to upper eyelid switch flap is designed and incised. Lower eyelid subciliary incision started all the way medially at the canthus and extended laterally up to the upper eyelid fold at the supratarsal crease. For the upper eyelid incision, a supratarsal fold incision is made and connects to the lower eyelid incision. The superior upper eyelid incision is made from the supratarsal fold point medially then carried up and lateral to the point of the planned lateral canthus. This laterally based triangular lid-switched flap is then raised as a skin/(orbicularis oculi) muscle flap.
8. Lateral canthus is grasped with a single hook, released sharply with scissors, and secured with a figure-of-eight, permanent 2-0 nylon to a lateral orbit drill hole.
9. The transposed lid switch flap is secured with 5-0 plain gut sutures. Again, care is taken to advance the flap all the way to the medial canthus region.
10. Coronal incision is closed in two layers.

Adolescence

After facial skeletal maturity (ranging from ages 14 to 18 years), correction of residual jaw abnormalities using orthognathic techniques can be safely performed without fear of continued growth disturbances (35,36). Presurgical orthodontics are essential to remove dental compensations in preparation for orthognathic surgery. Typically to correct an anterior open bite, often present in Treacher–Collins patients, a Le Fort I posterior impaction, a bilateral sagittal split osteotomy with advancement and a sliding osseous genioplasty is used (Fig. 3). However, the maxilla may be short and a posterior impaction may be difficult. Surgical planning typically involves lateral cephalometrics and fabrication of intermediate and final splints.

Finally, Tessier has described l'integrale procedure combining a Le Fort II osteotomy, bilateral mandibular advancement, orbital bone grafting, and genioplasty for the severe uncorrected form of Treacher–Collins syndrome with airway compromise. The series presented in 1986 reported correction of all 11 patients' reported respiratory symptoms (37).

For Treacher–Collins patients with persistent airway obstruction despite orthognathic correction into a class I occlusion, distraction of the genioplasty segment with a fascial sling, hyoid advancement has been described (38,39). The hyoid advancement improves the epiglottis position. The genioplasty distraction with gradual advancement is aimed at minimizing relapse from strong genioglossal muscle pull of the Treacher–Collins patient.



FIGURE 3 Preoperative (A,C) and postoperative (B,D) images of Treacher–Collins patient after zygomatic reconstruction with cranial bone graft, upper to lower eyelid switch flap, orthognathic correction (Le Fort I), bilateral sagittal split osteotomy of mandible, osseous genioplasty, and fat grafting. *Source:* Courtesy of Dr. H. K. Kawamoto.

Ancillary Procedures

The coloboma of the lower eyelid can be addressed after skeletal restoration of lateral midface projection and adequate orbital floor and rim support. This soft tissue problem similar to all aspects of the syndrome varies in severity. The coloboma affects all layers of the lid and reconstruction of mucosal lining, tarsus support, and skin closure are critical (9). Limited lateral defects are reconstructed with excision and resuspension of the lateral canthus. Larger defects are reconstructed with Z-plasty, preseptal orbicularis advancement, and tarso conjunctival wedge excision and closure (3,10). With large coloboma, reconstruction using upper lid to lower lid flaps combined with palatal grafting and fascial reconstruction of the tarsus are reasonable options for repair. Recurrence may be a problem.

Finally, soft tissue irregularities in the malar region associated with this condition can be improved with fat grafting for minimal to moderate deficiencies (3,10,40). Severe soft tissue hypoplasia requires free tissue transfer for improvement. We have also found that serial autologous fat transplantation during other childhood procedures has lessened the need for a large free flap procedure. Rhinoplasty often completes the soft tissue refinements needed after satisfactory skeletal correction. Del campo has described a procedure to narrow the base of the nose using rhinoplasty techniques (41). Resection of the broadened apical nasal pyramid followed by medial infracture has been coined the centrofacial flattening procedure.

SUMMARY

Treacher–Collins syndrome remains a difficult entity to treat often requiring multiple staged procedures throughout childhood. Despite difficulties with complete correction of this condition, improvements in functional status including early airway obstruction correction with distraction techniques and orthognathic surgery can make treating patients with this condition rewarding.

UCLA CRANIOFACIAL CLINIC PROTOCOL

The UCLA Craniofacial Clinic Protocol for correction of Treacher–Collins may vary depending on the individual patient and the severity of the deformity. However, guidelines for timing of the procedures are as follows:

1. Three months—Repair of cleft lip and/or one year: repair of cleft palate; if airway obstruction is problematic as an infant, surgically procedures may be used for correction of this including: a tracheostomy, mandibular or hyoid advancement or another procedures.
2. Six to eight years—Reconstruction of malar region—zygomatic arch, lateral orbital wall, and orbital floor with cranial bone graft and eyelid switch-flap correction.
3. Six to eight years—External ear reconstruction.
4. Five to eight years—Mandibular lengthening with distraction osteogenesis using intraoral devices.
5. 14 to 18 years (facial skeletal maturity)—Orthognathic (jaw) surgery.
6. 14 to 18 years (after jaw surgery)—Septorhinoplasty, laser removal of sideburn hair, or other ancillary (“finishing”) procedures.

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18

Craniofacial Tumors: Fibrous Dysplasia and Neurofibromatosis

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FIBROUS DYSPLASIA

Introduction

Fibrous dysplasia remains one of the most frequently encountered osseous craniofacial tumors (1). A benign fibro-osseous disease, its etiology continues to be poorly defined (2,3). Fibrous dysplasia was first described in publication by Von Recklinghausen in 1891 (4), and it was later fully characterized and named as such by Lichtenstein in 1938 (5). The pathologic process results in the gradual infiltration of normal bone with fibrous tissue intermingled with immature woven bone arranged in an erratic trabeculated pattern (6).

The most common type of fibrous dysplasia is the single bone variant, known as monostotic fibrous dysplasia. The polyostotic manifestation is four times less common than the monostotic variant and involves multiple skeletal sites (7). A more aggressive condition of polyostotic fibrous dysplasia, known as McCune–Albright syndrome, includes a classic triad of symptoms: (i) fibrous dysplasia, (ii) skin hyperpigmentation known as café-au-lait spots, and (iii) hyperendocrine states such as premature sexual development and hyperthyroidism. Patients with McCune–Albright syndrome may present with accelerated disease and increased morbidity when compared with other types of fibrous dysplasia (8).

The most frequently affected craniofacial bones are, in order, the maxilla, frontal, and then the sphenoid bone (5,9). However, the disease process frequently extends into adjacent bones, resulting in coincident involvement of the zygoma, orbital, and temporal bones (8). Although in its classic description, fibrous dysplasia was thought to “burn out” over time, there are many anecdotal reports that suggest that while extremity lesions may become inactive during adulthood, craniofacial involvement regularly progresses through adulthood (8).

Diagnosis and Presentation of Fibrous Dysplasia

The characteristic presentation of craniofacial fibrous dysplasia is a gradual, painless growth of the affected bone, resulting in facial asymmetry (5). Symptoms may develop as dysplastic bone encroaches upon nerves, sinuses, or periorbital structures. Optic nerve compression, trigeminal neuralgia, extraocular muscle palsy, and tinnitus or deafness have all been reported (10–14). Sinusitis or epiphora can occur (15,16). Rarely, tumor progression may result in airway obstruction. Not infrequently, the initial diagnosis is made incidentally on radiographs as part of the workup for these symptoms (5).

The differential diagnosis of fibrous dysplasia includes true bone tumors, such as ossifying fibromas, as well as giant cell granulomas or aneurysmal bone cysts (5,17,18). Cherubism, which is sometimes confusingly termed familial fibrous dysplasia, is a benign autosomal dominant giant cell lesion of the maxilla and mandible. Cherubism begins as bilateral painless growth of the jaws at two to five years of age, continuing until puberty, when the swelling regresses spontaneously, leaving no malformation (5,7,19,20).

Over time, fibrous dysplasia may result in sarcomatous degeneration. Currently, rates have been estimated to be 1% per year (21). Sarcomatous degeneration typically presents as a rapidly increasing mass within existing fibrous dysplasia (8). On computed tomography (CT) scan, this can appear as a new soft tissue lesion that is eroding into normal bony cortex (8,22).

Medical Management of Fibrous Dysplasia

Therapeutic medical treatment for fibrous dysplasia remains limited. Historically, radiation was used in an attempt to treat diseased areas, resulting in an increased frequency of sarcomatous degeneration. As a result, radiation is contraindicated as therapy for fibrous dysplasia. Researchers have noted that fibrous dysplasia results in increased cellular bone resorption activity; as a result, trials of bisphosphonates, which inhibit osteoclastic bone resorption, have been attempted with minor success. Published studies have reported a decrease in symptomatic pain and a reduction in molecular indicators of bone turnover (23–27). One significant drawback to these studies includes the lack of control groups (8). In addition, the craniofacial manifestations of fibrous dysplasia do not appear to be significantly affected by bisphosphonates (26).

Recommendations for continued medical management of slowly growing fibrous dysplasia include yearly monitoring at a multidisciplinary clinic (28). A neuro-ophthalmologic examination is necessary to document any early visual changes. Similarly, a complete audiologic assessment should be performed. A noncontrast CT scan is indicated if a new symptomatic or cystic lesion is noted, as this may represent sarcomatous degeneration (7). Controversy exists whether puberty or pregnancy may exacerbate fibrous dysplasia (29–31). While fibrous dysplasia tumors are known to be as hormonally responsive as normal bone, anecdotal evidence exists to suggest that physiologic hormonal conditions such as pregnancy do not worsen fibrous dysplasia (8). However, endocrinologic workup is indicated in all patients, since pathophysiologic hormonal states such as thyrotoxicosis or hyperparathyroidism are known to aggravate fibrous dysplasia (8,32).

Surgical Treatment of Fibrous Dysplasia

Due to the unpredictable course of fibrous dysplasia, the timing of surgical intervention in this disorder remains controversial. Advocates of a more aggressive approach suggest that early treatment with radical resection of diseased bone may avoid later damage, particularly in regard to vision. Prophylactic optic canal decompression, especially if there is extensive sphenoid involvement, may reduce compressive forces and protect the optic nerve (14,33,34). Others support a more conservative surgical method, contending that trauma from the surgery itself may cause increased edema surrounding the optic nerve, resulting in visual loss (6). These surgeons recommend less invasive recontouring, shaving, and curettage as surgical techniques of choice (35). Radical resection should be used only as a last resort, and optic nerve decompression should only be performed after documented evidence of visual loss (13,36).

Regardless of the timing of surgery, the objectives should remain unchanged: (i) maintenance of function, including airway, vision, hearing, and oral ability and (ii) improvement in appearance with correction of the disfigured bony structures. Modern craniofacial surgical techniques greatly improve the chances of success. Surgical oncologic tenets continue to hold true for planning: (i) precise preoperative imaging, (ii) appropriate surgical exposure, (iii) protection of critical structures, (iv) restoration of vital protective areas, including the dura and the great vessels, and (v) obliteration of dead space (6,37).

Every consideration should be made to completely excise dysplastic bone and to utilize normal bone grafts to reconstruct the resultant defect; however, in many cases, the extensive nature of the disease precludes total removal of the lesion and outstrips the supply of normal bone available for grafting. Edgerton and others have utilized fibrous dysplasia bone as grafts for reconstruction (35). Multiple treatment processes, including autoclaving and cryotherapy, have been used to obliterate the cellular elements, while maintaining the mineralized matrix (7,38–40). There are indications that such treated bone grafts demonstrate eventual substitution by normal bony architecture (40). The drawbacks of utilizing treated fibrous dysplasia bone are

that unpredictable resorption occurs, and that there is limited or no further growth of the grafted bone. This precludes its use in children (41). The fibrous dysplasia bone itself is also quite spongy and elastic, resulting in difficulty with adequate fixation (6).

Chen and Noordhoff approached surgical management of craniofacial fibrous dysplasia from an anatomic classification, dividing surgical sites into four “zones” (42). Zone 1, the fronto-orbital, nasoethmoid, and zygomatic areas, is intimately involved in the senses of smell and vision. As a result, the authors recommend complete excision of tumor with primary reconstruction with bone grafts (42). A fronto-orbital approach permits entry through the orbital roof, while an extradural and intradural approach allows complete visualization of optic nerve fibers (8). Zone 2, comprised of the cranium covered by hair-bearing scalp, is deemed less aesthetically critical. Recontouring by shaving and curettage is recommended. Zone 3, the temporal bone, is considered a difficult area to surgically treat due to the presence of large vessels and multiple cranial nerves. The functional deficits seen in temporal lesions include hearing loss and facial nerve palsies (8). Lastly, Zone 4 includes the tooth-bearing bones—the maxilla and the mandible. Orthognathic surgical techniques make aggressive resections more feasible than in the past. Modern oncologic mandibular reconstruction techniques utilizing free tissue transfer for hemi-mandible or subtotal mandible resections can result in excellent functional outcomes (8).

NEUROFIBROMATOSIS

Introduction

Neurofibromatosis originates from neuroectodermal tissue. As benign tumors, neurofibromas infiltrate skin, subcutaneous tissue, and bone (7). Growth is slow and irregular, but progresses inexorably. Neurofibromas have the potential to degenerate into soft tissue sarcomas. Although neurofibromatosis has an autosomal dominant method of transmission, 50% of all cases are thought to be spontaneous new mutations (43).

The most common type of neurofibromatosis is neurofibromatosis type 1, or NF1. Sometimes termed peripheral neurofibromatosis or Von Recklinghausen disease, NF1 accounts for 90% of all neurofibromatosis cases (44). Overall incidence of NF1 is 1 in 3500 live births. Cutaneous manifestations present in NF1 include neurofibromas, plexiform neuromas, and café-au-lait spots (45). The specific gene responsible for NF1 is located on the long arm of chromosome 17.

Neurofibromatosis type 2, or NF2, is much more rare, with an incidence of 1 in 210,000. Termed bilateral acoustic neurofibromatosis, the hallmark of NF2 is vestibular schwannomas. Other manifestations of NF2 are predominantly intracranial tumors and related spinal cord tumors (46). The pathophysiology is generally more severe in NF2 than NF1, and NF2 results from a different genetic mutation located on the long arm of chromosome 22 (45,46). Cutaneous and craniofacial manifestations are rare in NF2.

The most frequent symptomatic craniofacial manifestations of neurofibromatosis include sphenoid wing deficiency, with either hypoplasia or its complete absence. This results in a posterior orbital wall defect which allows the temporal lobe to herniate into the orbital cavity, producing a proptosis with associated globe pulsation (7,47). Conversely, enophthalmos can also occur if the interior orbital fissure becomes enlarged, or if the orbital rims and zygoma become hypoplastic due to disease involvement (7). Jackson classified orbital lesions into three categories: (i) orbital soft tissue involvement with intact visual function; (ii) orbital soft tissue and bone involvement with intact vision; and (iii) orbital soft tissue and bone involvement without residual visual function (48).

Medical Management of Neurofibromatosis

Annual neurologic, ophthalmologic, and dermatologic examinations should be conducted (49,50). In addition, an initial magnetic resonance imaging (MRI) of the brain should be obtained for any newly diagnosed child, or if additional symptomatology arises, including neurologic deficits, visual loss, or endocrinopathy, such as growth delay or precocious puberty (49,50).

Surgical Treatment of Neurofibromatosis

Most surgical experiences with neurofibromatosis are less than satisfactory. Treatment generally does not result in complete correction, and the disease itself continues to progress after surgery. Experienced authors highlight the high complication rate and the relatively modest improvement in appearance (51).

Recommended techniques for surgical treatment of neurofibromatosis including staging corrective procedures, including secondary eyelid adjustments, for maximum aesthetic improvement (44). A transcranial approach is recommended for larger orbital bony defects (44). If the orbital dystopia is severe, bony translocation, such as a box osteotomy, may be indicated. Zygomatic osteotomies can be used to reposition the orbital floor and reduce orbital apertures. Severely affected or blind globes may do best if enucleated and replaced with a prosthesis (44,52).

Difficulties typically encountered in surgical correction of neurofibromatosis include resorption of bone grafts with recurrent globe pulsation (44,53). Some reasons postulated for bone graft resorption include mechanical forces due to temporal lobe pressure, direct tumor erosion into bone grafts, or environmental factors such as abnormal dura that may be associated with the neurofibromatosis (44). Consequently, multiple authors have advocated utilizing alloplastic material for reconstruction. Titanium or vitallium mesh has been used successfully (44,52). Similarly, surgeons have reported good outcomes with custom stereolithographic porous methylmethacrylate implants designed from CT scans (54).

Similar to treatment of fibrous dysplasia, the timing of surgery to treat neurofibromatosis remains controversial. The decisions to intervene surgically requires an individualized approach for each patient and long-term evaluation both pre- and postsurgically to optimize success (44).

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19 Management of Facial Fractures

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INTRODUCTION

Facial fracture treatment ranges from the straightforward to the very complex. In all cases, however, the goal is to recreate the preinjury appearance and function of the patient's facial structures. The surgical approach is guided by the physician's clinical judgment that individualizes treatment according to the severity of the fracture and associated injuries. The definitive repair is carried out in one operative procedure, which will determine the degree of success or perfection achieved. Therefore, careful evaluation, diagnosis, and preoperative planning are critical to achieve the best surgical results.

The development of modern craniofacial surgical principles and techniques has resulted in dramatic improvements in facial fracture surgery. The best results are achieved by a multidisciplinary approach to the overall management of the injury. The trauma that causes facial fractures may involve injuries to a multitude of organs and structures that require close cooperation between all the physicians involved in a patient's care. The multidisciplinary team may involve a number of specialists including a plastic surgeon, ophthalmologist, neurosurgeon, otolaryngologist, oral surgeon, internist, anesthesiologist, orthopedic surgeon, orthodontist, and/or dentist. Patients with temporary or permanent alterations in appearance or function may require the assistance of "psychiatry," occupational, and/or physical therapy, prosthetics and potentially a cosmetician familiar with trauma. The overall goal is to return the patient to their preinjury state of function and appearance as soon as possible.

EVALUATION

By accepting the care of a facial fracture patient, you become responsible for the patient's overall care, therefore, a thorough evaluation of the patient's injuries is essential.

During the evaluation phase, the following needs to be determined:

- Location and extent of all injuries
- Facial structures injured
- Soft tissue loss (skin, mucosa, muscle, and nerve)
- Extent of bone loss
- Presence of dentoalveolar injuries

The initial consultation to evaluate a patient with facial trauma follows a systematic approach (1). A high index of suspicion for other injuries should be maintained throughout the evaluation process. As part of the evaluation process, the physician should obtain the mechanism of injury, the time of the injury, and any prior treatment the patient received. The assessment begins with the standard trauma protocol evaluating the airway, breathing, circulation, CNS status, and the cervical spine (2). Depending on when the evaluation is conducted, findings can change over time due to swelling, bleeding, or delayed effects of injuries to other organs or structures. After the initial survey, a more detailed physical exam is performed to exclude any problems that may not have been identified during prior exams.

Since there are often concomitant coexisting injuries to the cervical spine associated with facial trauma, a high degree of suspicion is required until the physician rules out that

possibility by exam or radiographs. If the patient has altered consciousness, whether from injury or medications, radiograph confirmation is strongly recommended.

Airway evaluation is an ongoing process. The airway may be compromised as a result of swelling or fracture displacement. Airway management becomes an issue depending on how the physician manages the fracture. A tracheotomy may be required to treat a compromised airway. The decision to perform a tracheotomy may be motivated by the need to avoid the physical presence of an endotracheal tube in the operative field during surgery.

If a patient presents with altered mental status, a high index of suspicion of intracranial injury should be maintained and immediate evaluation begun. In older patients, the assumption or previous history of altered mental status (i.e., dementia) should not divert one's attention. These patients require immediate evaluation to exclude any emergent intracranial trauma. Because of the potential of delayed effects in patients taking medications such as anticoagulation drugs, the observation of a patient's mental status requires ongoing evaluation. In cases, which are not clear, immediate consultation with a neurosurgeon is mandatory.

A thorough eye exam should be performed including visual acuity, inspection of the anterior chamber, retina, pupillary reflexes, and extra-ocular movement. An ophthalmologic consultation is advised. Difficulty occurs when a patient cannot cooperate to determine visual acuity. The uncooperative, sedated, or obtunded patient exemplifies this situation. Clinical assessment of the pupillary reactivity to light is essential. The evaluation for an afferent defect is performed using the "swinging flashlight" test (3,4). If there is a partial or total loss of vision in one eye, the opposite pupil will not react consensually. The test is performed in a dimly lit area. As the examiner moves the flashlight from one eye to the other, both pupils will dilate symmetrically when the light is directed into the non-seeing or poorly seeing eye. Both pupils will constrict symmetrically when the light is directed into the normal eye. A sluggish pupil in an uncooperative patient should be documented. Treatment, with steroids and serial exams of the pupils, should be initiated until a full evaluation can be completed. The patient's preinjury visual history can provide helpful information in assessing eye trauma. Failure to appreciate difficulties in vision or extraocular movements may be interpreted as a postoperative complication.

Examination of a patient's injuries begins with an overall inspection of the face for asymmetry, contusions, swelling, and lacerations. Palpation of the entire facial skeleton should note areas of irregularity or instability. The canthal attachments should be tested for stability, especially in cases of mid-facial trauma. The nose should be palpated and inspected for signs of fracture. Mandibular range of motion should be measured and the jaw joints should be examined by palpation. The oral cavity should be evaluated for malocclusion and obvious signs of dental trauma. If a tooth has been avulsed intact, replacement into the socket, using appropriate stabilization, should be considered. A chest radiograph is helpful in evaluating the presence of aspirated teeth or tooth fragments. The motor and sensory nerve function of the face should be evaluated.

When the physical examination is complete and all emergent issues stabilized, the fractures are evaluated radiographically. High-resolution computed tomography (CT) scans are currently the imaging procedure of choice for facial fractures (5–7). Ideally, high-speed, helical CT scans of the affected facial areas are obtained to delineate the anatomic fracture pattern. The CT scans, with 0.5 to 1.0 mm square pixels, can be formatted in axial, coronal, and sagittal planes (Fig. 1A,B,C). Advanced CT workstations can generate detailed three-dimensional reconstructions, which can be valuable in understanding the facial fracture (Fig. 1D) (8). CT scans can be extended to the cervical spine and cranium to exclude trauma. Panoramic tomography (panorex) may provide additional helpful information, especially in regard to the dentition and jaw joints.

While obtaining the patient's history, it is beneficial if recent close-up photographs are available, both profile and frontal. Even a driver's license photo may be helpful in evaluating the patient's preinjury appearance. A patient's dentures, whether partial or complete, may be helpful with fracture management. When treating adolescents and young adults, it is important to inquire about recent orthodontic treatment or retainers.

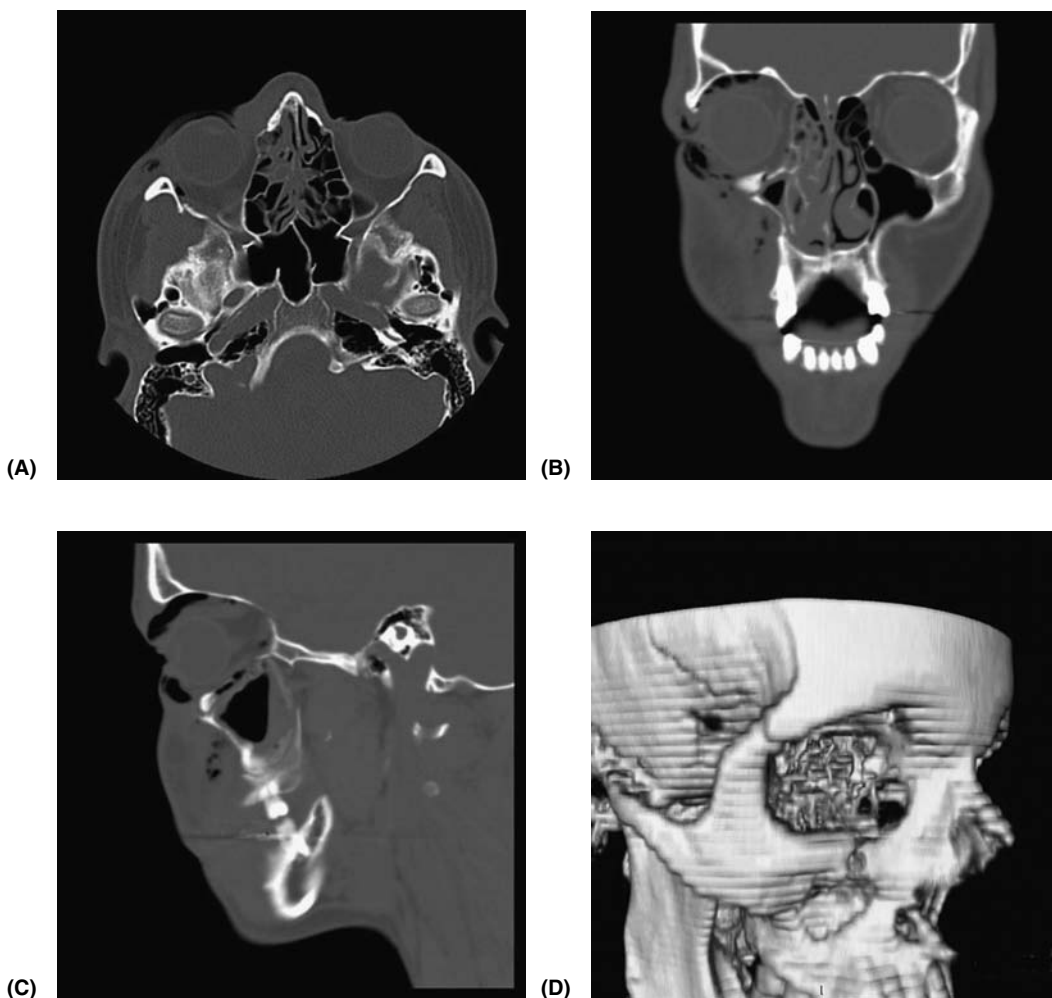


FIGURE 1 Computed tomography (CT) (A) axial, (B) coronal, and (C) sagittal images. (D) Three-dimensional CT reconstruction.

DIAGNOSIS/PLANNING

Once the history, physical exam, radiographs, and consultations are obtained, a definitive list of injuries can be made. A systematic approach to treatment requires that clinical judgment be the critical factor in regard to the indication and timing of surgery.

Analysis focuses on the following:

1. Exposure of the involved craniofacial skeleton
2. Fracture mobilization and accurate reduction
3. Methods of rigid bony fixation
4. Need for primary bone grafting and source of bone
5. Repair of soft tissues and suspension of the soft tissues to the skeleton

Incisions to expose the fracture sites should be determined preoperatively and, if possible, reviewed with the patient. Incisions may involve the use of existing lacerations, scalp incisions, periorbital incisions, and incisions in the oral cavity. The choice of incisions will be a clinical decision based on the surgeon's experience and clinical judgment.

The method of fracture mobilization to achieve reduction should be reviewed. Accurate and confident fracture reduction is critical for success. Adequate exposure is necessary to evaluate the reduction and to place the rigid fixation. The type of rigid fixation to use should be determined preoperatively and be available during surgery.

Airway management, both during and after the operation, should be analyzed with input from the anesthesiologist. Oral or nasal intubations are options as long as they do not compromise the surgery. Associated injuries (c-spine or basilar skull fractures) prevent their use. The need for a tracheotomy should be assessed during the initial evaluation of the patient and in preparation for the definitive repair.

INFORMED CONSENT

After developing a plan of action, a thorough discussion in regard to the potential risks of the operative procedure should be reviewed with the patient. If the patient is not able to make an informed consent, review these potential risks with the patient's family. The concerned parties should be aware that despite your best efforts, perfection should not be expected. Even in the most experienced hands, improper reduction of fractures can occur. Permanent scarring and residual signs of trauma are to be expected. Donor site problems, chronic pain, and infections that could be potentially life-threatening can occur. The possibility of additional surgeries to achieve the best results cannot be determined prior to treatment. The potential for loss of vision, intracranial injury, and even death are rare, but a reality, even in the best of care (9–11).

Unfortunately, the public's perception of what we are able to accomplish is based on highly edited television programs. Therefore, the need for education and informed consent cannot be emphasized enough. The fact that 30% of facial trauma patients have lawyers and are involved in some form of litigation should reinforce the importance of clear communication and appropriate documentation, using photographs and illustrations (11).

SURGERY SCHEDULING

In the treatment of a patient with multiple maxillofacial injuries, some injuries require immediate operative management, whereas, other procedures to treat other injuries can be delayed.

Immediate treatment in patients with maxillofacial injuries is indicated when the following are present:

- Airway compromise with severe maxillofacial trauma may require an immediate operation to temporarily reduce the fractured facial bones encroaching on the airway. Reducing the impacted fracture fragments and placing the patient into intermaxillary fixation can usually accomplish this. A tracheotomy may be necessary to control the airway and/or secretions. Control of the airway is critical.
- Severe hemorrhage from fractured bony segments may necessitate immediate surgery to ligate injured vessels or to reduce the segments and control hemorrhage. Large open wounds need to be debrided and closed early in a layered fashion.

Other maxillofacial injuries can be addressed immediately or up to two to three weeks later without significant compromise. It is prudent to delay surgery on unstable patients with intracranial injuries. Although once stabilized and even despite a low score on the Glasgow Coma Scale, early repair of facial fractures in these patients should be attempted, with input from neurosurgery (12). It is difficult to predict when an unconscious patient will improve. If the physician should choose to wait, the advantage of early repair may be lost, making subsequent attempts at repair exceedingly difficult, if not impossible (13).

FRONTAL SINUS FRACTURES

Anatomy

Understanding the anatomy of the frontal sinus is critical for the evaluation and treatment of injuries to this area (14,15). The frontal sinus is strategically located between the anterior cranial fossa and the nasoorbitoethmoid region in the central portion of the frontal bone. The frontal sinus is a pyramidal air-filled cavity lined by mucosa. It is located above the nose and makes up a good portion of the roof of the orbit. The size and shape of the sinus may vary. The thick anterior wall provides structural support. The posterior wall is thinner and separates the sinus from the frontal lobes of the brain. Sinus drainage is through the frontonasal ducts, bilaterally. The ducts are located in the anterosuperior part of the anterior ethmoid complex and exits usually into the middle meatus. Injury to the frontonasal ducts prevents drainage of normal mucosal secretions, which can cause a mucocele and/or infectious intracranial complications.



Evaluation

Clinical signs of frontal sinus injury may include overlying skin lacerations, bruises, hematoma formation, decreased sensation in the supraorbital nerve distribution, and cerebrospinal fluid (CSF) rhinorrhea. Physical examination consists of a thorough palpation of the forehead. Lacerations directly over the frontal bone should be carefully probed to assess anterior wall integrity. If a frontal sinus fracture is suspected, the nose must be carefully inspected for CSF. Any clear fluid should be sent to the laboratory for glucose determination (glucose > 30 mg/dL confirms CSF) or to check for beta-2 transferrin (16,17). Bloody nasal fluid may be tested at the bedside for CSF by placing a drop on a piece of tissue paper. Any CSF will diffuse faster than the blood and create a clear halo around a blood stain ("halo test") (18). Mental status, vision, and sense of smell should be documented. Eye injuries are common in patients with frontal sinus fractures. Therefore, an ophthalmologic evaluation is recommended.

Radiological evaluation of the frontal sinus is critical in determining the extent of the injury and the need for surgery. CT scan is standard in evaluating frontal sinus trauma. Scans should be performed with narrow cuts (0.5–1.0 mm) and include axial and coronal views with complete visualization of the frontal sinus, orbits, and nasoethmoidal areas. Evaluation of the integrity of the anterior and posterior sinus walls and frontonasal duct system is crucial in developing a treatment plan. In many instances the frontonasal ducts are difficult to assess. Isolated fractures of the anterior wall of the frontal sinus, and transverse linear fractures of the anterior and posterior walls above the floor of the sinus are not usually associated with damage to the frontonasal ducts. Fractures involving the floor of the sinus, the nasoethmoid complex, or depressed fractures of the posterior wall almost always signify injury to the frontal sinus drainage system. CT scan findings of a wide gap in the posterior wall or depressed fractures of the posterior table are usually associated with a torn dura (19).

Management

The goals of management are both to prevent a visible deformity and early and late complications (20–22). These include acute and chronic sinusitis, mucocele formation, meningitis, and brain abscess. All patients receive antibiotics tailored for upper respiratory bacterial flora at the start of treatment and for seven days after surgery, and 14 days after surgery if a CSF leak is stopped.

Careful analysis will determine the minimum operative intervention necessary to achieve the best result. Knowledge of the specific wall involvement, whether the frontonasal duct is involved, or whether there is a CSF leak are all important considerations. Sometimes this cannot be determined without operative exploration. If a fracture of the anterior table occurs with no

displacement, surgery is not needed (23). If the anterior wall is displaced, open reduction and internal fixation best treats the fracture and avoids a frontal bone aesthetic deformity.

The sinus may be approached through an appropriate skin laceration or with a coronal incision. The coronal incision provides the best exposure and should be elevated with the pericranium to be utilized as a separate flap, if needed. The sinus should be explored, irrigated, and all devitalized mucosa removed. If no frontonasal duct injuries are present, the healthy mucosa is left intact and the anterior sinus wall is anatomically reduced and fixated with an absorbable or metallic microplating system. When plating the anterior sinus wall, close attention to the convexity of the bone is especially important in the brow area. A common error is to reduce and plate this area in a "flat" position. If significant bone loss is present or if fragments are judged to be too small to use, split calvarial grafts or "shavings" from the temporoparietal area are used to fill the defects.

Any fracture that extends into the floor of the frontonasal duct may produce obstruction and therefore requires sinus mucosa ablation and frontonasal duct obliteration (24–26). If desired, intraoperative evaluation of frontonasal duct function can be assessed by attempting to pass a liquid through the ducts (fluorescein or methylene blue) (27). If the dye does not pass into the nose, the ducts are obstructed. Any evidence of frontonasal duct obstruction makes complete removal of the sinus mucosa and obliteration of the sinus and ducts mandatory. This is performed by carefully removing all the mucosa from the sinus and burring all the bony surfaces with a high-speed round diamond burr. The burr removes a thin layer of bone from the sinus. The frontal sinus mucosa invaginates within small recesses in the bone on the inside of the frontal sinus.

A pericranial flap can be elevated from the coronal flap, or from the cranium, and placed over the ducts with bone grafts to plug the duct system (28,29). Obliteration of the frontal sinus is recommended. The best material for frontal sinus obliteration remains debatable (30–32). Our preference is to use calvarial bone grafts and a pericranial flap (Fig. 2). If flaps are used, an opening in the lower aspect of the anterior sinus wall should be left open so that the pedicle is not compressed. Spontaneous osteogenesis, galeal-frontalis flap, fat and muscle are other options (33,34). Spontaneous osteogenesis of the frontal sinus occurs by the slow in growth of bone from the burred down inner cortex. This technique avoids a donor site. The size of the frontal sinus varies among individuals and with the variety of options available, the surgeon's experience and judgment must be used as the determining factor. If one of the frontonasal ducts is injured, we prefer bilateral duct and sinus obliteration unless a complete septum is present and the non-injured sinus is pristine.

Fractures of the posterior wall of the frontal sinus are often associated with intracranial injuries. CSF leak and frontal lobe contusion may be present. Evaluation and treatment should involve neurosurgery. Management is based on the degree of displacement, duct involvement, and the presence of a CSF leak. No operative intervention is required for posterior wall fractures that are not displaced and exhibit no evidence of a CSF leak. All fractures of the posterior sinus wall that are significantly displaced require operative intervention. In the absence of CSF leaks and frontonasal duct injuries, the displaced frontal sinus fractures can

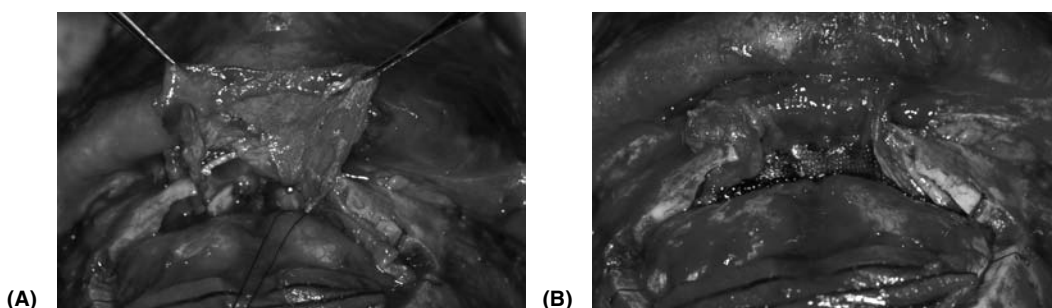


FIGURE 2 (A) Elevated pericranial flap above the naso-orbito-ethmoidal area. (B) Pericranial flap covering ducts and bone grafts during cranialization procedure.

be reconstructed with rigid fixation of both the anterior and posterior walls. If there is no CSF leak but the frontonasal ducts are injured, complete mucosal removal, with sinus and duct obliteration, should be performed. Displaced fractures of the posterior wall associated with a CSF leak require a cranialization procedure. Cranialization is the removal of the posterior wall of the frontal sinus, which allows the brain to displace anteriorly. After the posterior wall is removed, the dural injury can be repaired. Complete mucosal ablation on the remaining frontal sinus bone is followed by obliteration of the frontonasal ducts with bone grafts as described above. A pedicled pericranial flap may be obtained and advanced over the ducts, providing a vascularized barrier that can also be used for dural repair. The galeal-frontalis flap based on the supratrochlear and/or the supraorbital vessels, which provides greater bulk and vascularity, is another option. The anterior sinus wall is reduced and rigidly fixed. CSF leaks with nondisplaced or minimally displaced fractures are rare. These fractures are observed for five to seven days to see if the leak resolves spontaneously. If the leak continues, cranialization of the frontal sinus and repair of the dural laceration is performed.

Complications

The most frequent complications after frontal sinus fracture repair are frontal headaches, sinus infections, drainage problems, and contour irregularities (34,35). Headaches are ill defined and if problematic, require a CT scan to determine whether a mechanical cause can be seen. If a mechanical cause is not seen, a consultation with a neurologist is recommended. Unrecognized or prolonged CSF leaks may predispose a patient to develop meningitis. CSF leaks are an open communication between the CSF and the contaminated upper respiratory tract. Early repair of the dural defect is the best method to avoid an infection. Incomplete mucosal removal may allow new mucosa to grow within the sinus, producing mucocoeles, which can become infected (mucopyocele). Over time, mucocoeles may produce symptoms of chronic sinusitis and eventually cause bony erosion of the walls and floor of the frontal sinus. Symptoms include fever, pain, headache, and ocular pain. A CT scan is the most effective method of evaluation. Frontal sinusitis is initially treated with broad-spectrum antibiotics and decongestants. If medical treatment is unresponsive, transnasal endoscopy to facilitate drainage can be attempted. If this proves to be unsuccessful, surgical exploration and obliteration of the sinus is performed. Meningitis, brain abscess, or epidural abscesses require operative intervention. Treatment involves drainage of the abscess, repair of the dural tear, and cranialization of the frontal sinus.

Frontal sinus fracture patients should be followed annually for at least five years. Complications may not develop for many years after the injury and this fact must be emphasized to patients. Any time a patient with a history of frontal sinus fracture complains of frontal pain, swelling, pressure, visual changes, unexplained fevers, or persistent frontal headaches they should be evaluated clinically and radiographically (CT scan).

NASAL FRACTURES

Anatomy

The nose is the most prominent facial feature and therefore the most likely to be fractured. Nasal fractures are common injuries and are often considered a minor injury (36). Contrary to this perception though is the high incidence of posttraumatic nasal deformities requiring secondary treatment (37,38). The paired nasal bones articulate in the midline with each other, laterally to the frontal process of the maxilla, and superiorly to the nasal process of the frontal bone. The thinner, caudal portion of the nasal bones articulates with the upper lateral cartilages. This area is vulnerable to fracture and dislocation. The cartilaginous septum is quadrangular in shape and located between the perpendicular plate of the ethmoid and the vomer. The hard palate is the floor and the cribriform plate is the roof of the nasal cavity.



Evaluation

A detailed history and physical examination are essential for diagnosis and treatment. A precise assessment of the injury is recorded, including the mechanism of injury, direction of the blow, and the time of the nasal injury. Some patients may have difficulty communicating their preinjury nasal shape. A review of old photographs or a driver's license can be very helpful. Instruct the patient and family that multiple views are helpful, the less flattering the better. The physical examination of the nose includes inspection for lacerations, swelling, and deviation. The affected area is examined by palpation to identify tenderness, crepitus, depression, nasal shortening, or widening of the nasal base. An internal examination is performed with particular attention to the posterior septum; looking for signs of fracture and obstruction. The internal evaluation is performed with an endoscope that can be used to examine the inferior meatus, turbinates, septum, and the posteroinferior septal junction with the perpendicular plate of the ethmoid (39). If the posterior septal region cannot be visualized or if the force of injury and examination suggest an associated nasoorbitoethmoid fracture, a CT scan is obtained.

Management

Prompt diagnosis and treatment of septal hematomas are essential to reduce the incidence of fibrosis with subsequent septal distortion, abscess, necrosis, and saddle nose deformity. Wide dependent drainage followed by careful packing with antibiotic gauze and systemic antibiotic coverage is recommended. Small hematomas can be aspirated and followed closely for recurrence.

The septum is the key to nasal fracture management. If there is significant posttraumatic swelling, making precise reduction difficult, the patient is instructed to elevate the head and use ice with follow-up in three to five days. Definitive treatment is instituted five to seven days after injury, depending on edema resolution. If significant septal fractures are visualized, reconstruction by directly repositioning the septum is considered.

The reduction of the nasal bones is initially accomplished by recreating the fracture. Impacted nasal bone fractures require instrumentation to restore nasal length. If the nasal septum is dislocated, begin by relocating it into the vomerine groove. After reducing a comminuted nasal fracture, a dorsal posterior intranasal pack of absorbable gelatin sponge (gelfoam) can be used to prevent collapse after reduction.

A non-reducible posteroinferior or anterior septal fracture is a consideration for acute septal reconstruction. A hemi-transfixion, or Killian incision, is made and bilateral inferior mucoperichondrial flaps are developed. The septum is completely visualized and reduced. Reduced fractures of the septum are approximated with through-and-through (mucosa septum mucosa) chromic mattress sutures. Doyle splints are recommended to stabilize the caudal septum. Internal and external nasal splints are used for one week with prophylactic antibiotic coverage (40).

Complications

A septal hematoma may develop at the time of injury or after the repair. Untreated hematomas may become organized, causing fibrosis and obstruction. Hematomas under excessive pressure can cause necrosis with subsequent perforation. Loss of septal support can cause collapse of the cartilaginous dorsum resulting in a saddle nose deformity. Synechia can occur if lacerations in the septal and turbinate area are left in proximity to heal together. Slight malunion of nasal fractures is common after closed reduction and may require secondary reconstructive rhinoplasty.

NASOORBITOETHMOID FRACTURES

Anatomy

Nasoorbitoethmoid (NOE) fractures involve the area between the eyes. This type of fracture involves the bone to which the medial canthus is attached. The frontal processes of the maxilla

forms the principle vertical buttresses. The horizontal buttresses are the frontal bone and supraorbital rims superiorly and the infraorbital rims inferiorly. The thinner, delicate bones of the medial orbit include the lacrimal bones and lamina papyracea of the ethmoid bones. These bones fracture easily and can produce a medial wall blowout fracture. Fractures of the nasoorbitoethmoid complex displace the structures backward into the face. As a result, patients with NOE fractures have a flattened nose and swollen medial canthal areas. The interorbital space is located below the floor of the anterior cranial fossa and between the medial orbital walls. It contains the two delicate ethmoidal labyrinths divided by the central perpendicular plate of the ethmoid and nasal septum. The area between the eyes offers no significant structural support and when it fractures, the area readily collapses. These fractures can extend into the anterior cranial fossa adjacent to the cribriform plate with resultant CSF leakage. Frontal lobe contusion and olfactory nerve disruption can occur.



Evaluation

Soft tissue edema often obscures the physical findings associated with NOE fractures and a high degree of suspicion is necessary to confirm the diagnosis. Palpation of the medial orbital rim area can determine instability or displacement of the NOE fracture. Instability of the medial canthal attachment can be assessed by palpating the medial canthus while applying traction on the lateral canthus (bowstring test). A bimanual exam is performed by inserting a clamp intranasally into the area of the medial canthus while placing the index finger externally on the medial canthus. If there is movement, an unstable NOE fracture is present (41). A CT scan, both axial and coronal studies at intervals of 0.5 to 1.0 mm, remains the best imaging study for this area. Coronal studies can only be obtained after the cervical spine is cleared. Newer, spiral, high-speed CT scans offer exceptional axial, coronal, and sagittal views without manipulation of the cervical spine.

Management

The goal in the treatment of the NOE fractures is to restore the preinjury intercanthal distance, restore nasal projection and dorsal contour, and restore normal soft tissue contours in the medial canthal area. NOE fractures are the most challenging fractures to treat in facial trauma. Optimum surgical treatment involves wide exposure and meticulous reduction of bony fragments with adequate stabilization (42,43). All displaced NOE fractures require operative intervention. Any instability on NOE manual examination requires operative intervention. Fractures that do not show any movement on examination or displacement on the CT scan do not require surgery. Management of the medial orbital rim segment, which contains the canthal tendon, is critical in the successful treatment of these injuries.

Once the NOE fracture has been diagnosed and a CT scan obtained the fracture pattern and associated fractures should be identified (44). These two items determine the surgical incisions and type of exposure needed. For complete exposure of the NOE area, a coronal, lower eyelid, and gingivobuccal sulcus incisions are required. The most difficult area to expose is the medial orbital wall segments. Care must be taken when dissecting the medial wall fragments so that the medial canthal tendon is not stripped off the bone. The lower eyelid incision allows exposure of the inferior orbital rim and inferior medial orbital rims, as well as the orbital floor. A maxillary gingivobuccal sulcus incision is required to provide exposure of fractures involving the nasomaxillary buttress. The vertical midline incision on the nose is a good option in the elderly or bald patient with an isolated NOE fracture.

A Type I fracture involves a single segment, minimally displaced, with the medial canthus attached. This type of fracture can be reduced and managed by rigidly fixating the segment from below and above. The more common fracture patterns are Type II and Type III. Type II is a comminuted fracture, but the medial canthus remains attached to the bone (45). In Type III fractures the medial canthus is avulsed from the bone. These fractures are difficult to reduce and stabilize and a much wider exposure is necessary. Coronal, lower eyelid and maxillary gingivobuccal sulcus incisions are required. In the more comminuted fracture, identifying the bony fragment with the canthal attachment can be difficult. It may be necessary to make a small transverse incision over the medial canthus to identify the tendons and bone fragment to which they are attached. If necessary, the fractured nasal bones can be temporarily removed or displaced to provide better exposure of the medial orbital walls. This is an important maneuver because it provides excellent exposure for placement of transnasal wires.

Placement of transnasal wires with a parallel type reduction of the medial orbital rims is the key to obtain the best aesthetic results. This is conceptually easy to describe but in reality, technically difficult to perform (46). Perfection may be difficult to achieve. The medial canthal tendon insertion should be preserved. Transnasal wires (26 or 28 gauge) are passed through drill holes placed superiorly and posteriorly to the medial canthus on the central bone fragment. On larger bone fragments, an additional wire placed inferiorly may be used to prevent rotation. Tightening should be performed to a point of slight overcorrection. The remaining comminuted segments are reduced and stabilized with wires or small plates in the medial canthal area. If the fracture is unilateral, a wire-passing drill bit can be angled. However, in bilateral fractures it is technically difficult and requires the maximum exposure. In drilling the transnasal wires, minimum attempts should be used and each side done one at a time, with careful passage of the wires. If the medial canthal tendon is avulsed, the medial canthal tendon should be identified through a separate transverse incision overlying the tendon. A 3-0 wire suture is placed as a mattress suture into the tendon and attached to the central bone fragment or to a bone graft. Bone grafts are frequently used to reconstruct the nose and orbit in NOE fractures.

Lastly, the restoration of nasal projection and contour are critical to achieve the best result. The reduction and stabilization of nasal bones do not guarantee the return of the preinjury nasal appearance. The nasal septum should be carefully assessed on CT and clinical examination. Collapse of the nasal septum, with loss of support to the distal half of the nose, is frequently present in these injuries. If the tip of the nose can be pressed down to the nasal spine, septal support has been lost. Since reduction of the septum is often not adequate, a cantilever bone graft is needed to restore normal projection to the tip, and smooth the contour to the nasal dorsum. The full thickness outer table cranial bone graft from the temporoparietal region is the preferred source (Fig. 3). The cantilever bone graft can be secured with two screws superiorly to the frontal bone or stable nasal bone. The existing nasal bone may need to be burred if a preexisting hump exists so that the graft will lay flat and have better contact. The graft should be contoured and rounded to achieve the best aesthetic result with specific attention to the radix area so that this is not elevated causing a change in the frontonasal angle. The septum should be reduced and splinted with Doyle splints.

Redraping of the soft tissues is a crucial last step in the surgical correction of NOE fractures. The dissection necessary to reduce the bony fragments in this area results in an elevation of the soft tissues from the medial canthal area. If steps are not taken to ensure proper re-draping of the soft tissues, hematoma and scarring can result in the loss of the contour to this area. Padded nasal plates or bolsters are positioned over the nasoorbital valley and lateral nose to re-drape and assist in the re-adaptation of the skin to the underlying bone. Transnasal, pull-out wires can be placed to secure these splints or bolsters. The wires securing the splints or bolsters have no role in maintaining the intercanthal distance, but are effective in controlling soft tissue contour. Postoperatively, the bolsters should be inspected periodically to ensure that the underlying tissues remain healthy. The bolsters are removed after one week and the Doyle splints after two weeks. The patient is advised not to blow his or her nose and to avoid increasing intranasal pressure for two months after surgery.

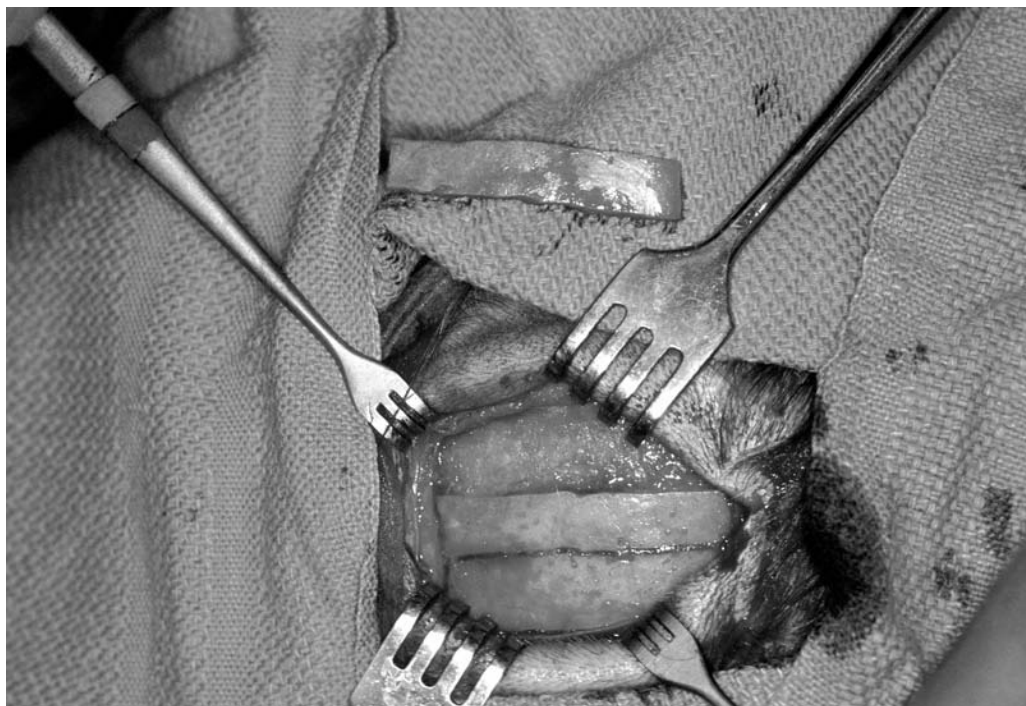


FIGURE 3 Full-thickness outer table cranial bone graft—the preferred source of bone in craniofacial reconstruction. *Source:* Courtesy of Dr. H. K. Kawamoto.

Complications

The nasoorbitoethmoid region is a delicate area. It is difficult, at best, to achieve the preinjury appearance. Some degree of malposition, whether of the bone and/or soft tissues, is common. Malunion, causing posttraumatic telecanthus, is characterized by an increased intercanthal distance. The occurrence of posttraumatic telecanthus correlates with the severity of the injury and may require secondary reconstruction. Disruption of the lacrimal system may occur. If symptomatic, a drainage procedure may be required. Routine exploration of the canaliculi is not recommended during fracture repair because the incidence of injury is low. The lacrimal system should be observed for several months and re-tested before deciding to perform a drainage procedure (dacryocystorhinostomy).

ORBITOZYGOMATIC FRACTURES

Anatomy

An orbitozygomatic fracture is another descriptive term for malar or tripod fractures. The “orbitozygomatic” term emphasizes the orbital component of the fracture pattern. The zygomatic bone gives prominence to the cheek. The zygoma articulates superiorly with the frontal bone, medially with the maxilla, laterally with the temporal bone (zygomatic arch), and posteriorly with the sphenoid. The zygoma is the lateral wall and floor of the orbit and articulates over a broad area with the sphenoid bone in the orbit.



Evaluation

The surgeon should have a good understanding of the important aspects of the physical examination, particularly with respect to the ophthalmologic findings. Clinical signs of an orbitozygomatic fracture include cheek and eyelid edema, loss of cheek projection, unilateral epistaxis, decreased or loss of sensation in the cheek area and upper lip, step off deformity in the infraorbital rim area, and diplopia due to displacement of the globe.

If present, swelling will conceal the degree of displacement and enophthalmos. Point tenderness and bony step offs should be assessed and correlated to radiographic findings. Sensory changes in the infraorbital nerve distribution should be evaluated and documented. The most important part of the preoperative surgical examination involves the results of the eye evaluations. Failure to identify difficulties in vision or extra-ocular motion may be interpreted as a postoperative complication.

Management of orbitozygomatic fractures requires the surgeon to accept responsibility to rapidly and accurately diagnose, and therapeutically intervene when acute conditions threaten the patient's vision following trauma or surgery. This is even more difficult when dealing with the uncooperative, sedated or obtunded patient, when accurate assessment of vision is difficult, if not impossible. The presence of a "Marcus Gunn" pupil (afferent pupillary defect) is indicative of optic nerve dysfunction. The best clinical test of optic nerve dysfunction is the "swinging flashlight" test. Even with a partial or total loss of vision in one eye, the pupils may appear normal in size and be equal in diameter on examination. In a dimly lit room, the examiner moves the flashlight from one eye to the other (47). Both pupils will dilate symmetrically when the light is directed into the non-seeing or poorly seeing eye, and both pupils will constrict symmetrically when the light is directed into the normal eye. On the other hand, a non-constricting pupil should direct the physician to assess the potential causes of cranial nerve III dysfunction, such as increased intracranial pressure from trauma. As a side note, the use of an epinephrine-based local anesthetic, which can directly stimulate the sympathetic pupillary muscles causing dilation, can be a common cause of intraoperative dilation of the pupil.

All patients in whom an orbitozygomatic fracture is suspected should have coronal and axial CT scans. Analysis of the scans should focus on the degree of displacement and internal orbital injuries. The lateral orbital wall provides an excellent area to evaluate the degree of displacement. The zygoma articulates with the greater wing of the sphenoid over a broad area that can be used in evaluating the degree of displacement. On the axial views, the zygomatic arch, zygomaticofrontal suture area, infraorbital rim, and zygomaticomaxillary buttress should be evaluated. The degree of displacement and comminution will be important in surgical treatment, and in deciding which incisions to use in addressing the fracture. Coronal CT scans are useful in evaluating the orbital component of orbitozygomatic fractures. This evaluation is important because defects of the floor or medial wall require reconstruction to prevent enophthalmos. The CT scan can confirm entrapment of the extraocular muscles that will require operative release. It is critical to remember that direct coronal views require the patient's neck to be hyperextended. If the cervical spine has not been cleared of injury, direct coronal views cannot be obtained. In these situations, reformatted axial views can be used to reconstruct coronal views. However, these views will be of an inferior quality compared to direct views. The reformatted images should still provide valuable information in evaluating the fracture pattern. When coronal reformatting is planned, it is helpful to obtain 0.5 to 1.0 mm spacing, which will provide better coronal reconstructed images. Newer, spiral, high-speed CT scans offer exceptional axial, coronal, and sagittal views without manipulation of the cervical spine.

Management

Orbitozygomatic fractures with no displacement require only observation. Patients should be cautioned against any potential pressure that may displace the fracture. If the fracture is displaced, exploration with anatomic reduction and plate fixation is indicated. Controversy exists regarding the amount of exposure and type of fixation used. The experience and clinical judgment of the surgeon will dictate the exposure and methods of fixation. Regardless of the

methods chosen, the anatomic reduction of the fracture is critical, followed by appropriate fixation (48–50).

In general, to restore the orbitozygomatic fracture, one should explore and properly align the zygomaticofrontal suture area, the infraorbital rim, and the zygomaticomaxillary buttress. The surgeon may choose to expose less and fixate fewer areas based upon experience and the confidence that the fracture is adequately reduced and fixated in position. The fractured zygomatic arch is an important component of orbitozygomatic fractures. The zygomatic arch is the key to facial width and projection (51,52). The vast majority of orbitozygomatic fractures do not require a coronal incision to expose and plate the arch. However, when the fracture sites are comminuted or if there is concern about the adequacy of reduction, exposure of the zygomatic arch will provide confirmation regarding adequate reduction (53). When exposed, the zygomatic arch should be plated first. When reducing the fracture, it must be remembered that the zygomatic arch is a relatively straight structure. A common error is to plate the arch with too much of a curve.

A gingivobuccal sulcus incision provides excellent exposure of the maxilla. On edentulous patients, the incision is placed on the alveolar ridge. Dissection should first be accomplished along the zygomaticomaxillary and nasomaxillary buttresses to the level of the infraorbital rim. At this point, a thick elevator can be placed under the distal arch and zygoma area. Constant strong pressure should be used to reposition the fracture from its impacted position. Anatomic reduction or even partial improvement will assist in approaching the infraorbital rim, since a displaced infraorbital rim may cause difficulty with the lower eyelid dissection. Next, the lower eyelid incision is made. The choice of which lower eyelid incision to use depends on the surgeon's experience (54–58). The trend has been to avoid transcutaneous approaches, especially the subciliary incision, due to the higher risk of lid retraction. The lower eyelid transconjunctival incision, with or without a canthotomy, is being favored due to the lower risk of lid retraction (59–62). Whatever approach is utilized should be performed carefully with a minimum of trauma to the tissues. The zygomaticofrontal suture is exposed by a separate incision in the lateral aspect of the supratarsal fold of the upper eyelid.

Lastly, for comminuted fractures or displaced fractures of the zygoma, a coronal incision may be needed to accurately reconstruct the facial width and projection by exposing the zygomatic arch. The greatest error is not adequately reducing the fracture. The key to this exposure is identifying the superficial layer of the deep temporal fascia, which is a glistening transparent layer overlying the superficial temporal fat pad superficial to the thick white deep temporal muscle fascia. The superficial layer of the temporal fascia becomes visible lateral to the orbital rim just below the level of the zygomaticofrontal suture (63). This fascial layer is incised and dissection continues through the fatty layer (superficial temporal fat pad), down to the zygomatic arch below the periosteum, avoiding injury to the frontal branch of the facial nerve.

The fracture fragment needs to be aggressively mobilized for proper reduction and fixation. Once the fracture is reduced, and if the zygomatic arch is not exposed, the first area to be stabilized is the zygomaticofrontal fracture using a wire to pass through two holes drilled laterally through the thick rim of the orbit. The wire is twisted and the wire knot is hidden beneath the lateral edge. A small fixation plate, using 1.0 to 1.5 mm screws, is another option. The fracture fragment can now be rotated and anatomically reduced. The zygomatic arch, if it is exposed, and the zygomaticomaxillary buttress should be plated with the largest plates for maximum stability using screws that measure at least 1.5 mm. The zygomatic arch, if it is exposed, is plated first. The zygomaticomaxillary buttress is plated last. The infraorbital rim can be treated with smaller plates and screws that are 1.0 to 1.5 mm in diameter. The inferior orbital rim must be held upward and forward while fixation is applied. The inferior orbital rim is level and not curved. Before placing the large plates, the inner lateral orbital wall can be visualized to confirm adequate reduction in the region of the zygoma and the greater wing of the sphenoid.

Once the zygomatic articulations have been plated, attention is focused on the orbital area. The orbital floor and medial wall are usually fractured. Indications for repair include defects large enough to cause enophthalmos and/or mechanical entrapment of the globe. A large variety of materials are used to rebuild these defects including bone grafts, metallic or absorbable mesh, and porous polyethylene (64,65). In grafting the orbital floor, the

posterior ledge of intact bone in the posterior orbit should be identified. One method uses a Freer elevator to slide up the posterior maxillary wall and feel the ledge. Another method uses an endoscope inserted into the maxillary sinus (66,67). Our preference is a cranial bone graft from the temporoparietal area with the pericranium attached. This graft is harvested with a wide sharp osteotome raising a “shaving” of bone and pericranium (Fig. 4). The elevated graft has a natural curvature making it perfect for this application. It should be remembered, that because of edema, the corrected side should be slightly overcorrected compared to the uninjured side. After the fracture has been treated, a “forced duction” test is performed by using toothed forceps to grasp the conjunctiva peripheral to the cornea to confirm unrestricted movement of the globe.

At the end of the procedure, attention should be focused on the soft tissue closure. The cheek tissue should be re-suspended to the infraorbital rim using sutures (68,69). The suture can be attached to a screw, the plate, or a hole drilled into the rim itself. A lateral canthopexy should be performed by reattaching the lateral canthus to the inner aspect of orbital rim periosteum, superiorly. Lastly, the periosteum should be closed over the zygomatic arch, the infraorbital rim, and the frontozygomatic area.



(A)



(B)

FIGURE 4 (A) Cranial bone graft “shaving”outlined in the temporoparietal region. (B) Bone shaving elevated with pericranium attached.

Complications

Diplopia is a known complication of an orbitozygomatic fracture (70). The complication can be caused by mechanical entrapment, contusion of the extraocular muscles, or the nerves supplying the muscles. A postoperative CT scan can rule out any concern that the muscle or periorbital area are entrapped. If no mechanical cause for diplopia can be found, the patient should be followed for three to six months. Extraocular muscle malfunctions usually resolve over time. If the diplopia should persist, extraocular muscle rebalancing should be considered.

Enophthalmos is a common, problematic complication (71–73). Enophthalmos may be caused by malposition of the zygoma, inadequate reconstruction of the internal orbit, orbital fat atrophy, loss of ligamental support, or a combination of these conditions. When enophthalmos is seen, a postoperative CT scan of the orbits should be obtained. Problems tend to occur because grafts or materials used to reconstruct the floor are placed too low, and not on the intact posterior ledge. Treatment may include bony reconstruction of the walls and floor of the orbit, or an osteotomy and repositioning of the fractured areas, or a combination of these.

Sensory changes due to the injury of the infraorbital nerve will cause sensory changes in the cheek and upper lip areas. Permanent hypesthesia is a possibility (74). Usually the paresthesias will resolve over time. If problems continue, a CT scan can be obtained to rule out any mechanical causes. If any are present, these mechanical causes can be treated by surgical decompression.

Infections are rare, but do occur, especially when alloplastic implants are used. Infections may require debridement and removal of loose hardware, implants, and necrotic tissue.

Lid retraction, following a lower eyelid incision, is a troubling complication. Lower lid retraction, especially with the use of subciliary transcutaneous incisions, can result in ectropion. This may resolve over several months with massage and taping, or it may remain a problem and require surgical correction.

Blindness after facial fracture surgery is very rare (75). Visual acuity and/or pupillary reaction should be tested as soon as possible after surgery. Irreversible ischemic damage often occurs within the first one to two hours after surgery (76). Management of post-surgical vision loss must be treated immediately with surgical decompression and high dose steroids. Begin with an intravenous loading dose of methylprednisolone (30 mg/kg). Two hours later, the methylprednisolone is given in 15 mg/kg increments intravenously. This dosage is continued every six hours (77). After decompression, the vision should be monitored and a CT scan performed to ensure adequate decompression. Despite treatment, prognosis is poor.

ORBITAL FRACTURE

Anatomy

Seven bones make up the orbit: the frontal, maxilla, zygoma, lacrimal, ethmoid, greater wing of the sphenoid, lesser wing of the sphenoid, and the palatine bone. The first four constitute the outer rim of the orbit and protect the more delicate bones that constitute the walls of the interior orbit. Immediately behind the infraorbital rim the floor is concave, while further back it becomes convex. The medial orbital wall is made up of the lacrimal bone and the thin lamina papyracea of the ethmoid, which is susceptible to fracture.

The floor of the orbit is the shortest of all the walls, extending back 35 to 40 mm from the infraorbital rim. The surface of the orbital floor is triangular and extends from the maxillary ethmoidal articulation to the inferior orbital fissure, and from the orbital rim back to the posterior wall of the maxillary sinus.



Evaluation

Initial evaluation includes a test of visual acuity, extraocular muscle function, and pupillary reflexes. Serial examinations are performed throughout the course of treatment. In the patient who cannot cooperate, pupillary responses should be examined.

The main signs of a blowout fracture are diplopia and enophthalmos. Restricted ocular movement can cause diplopia, most commonly in upward gaze. Enophthalmos, or the posterior displacement of the globe, is caused by increased orbital volume and/or a disruption of the ligamentous support of the globe. A thorough ophthalmologic evaluation should be performed prior to repair. If optic nerve injury or complete blindness is present, treatment should be started with high dose steroids while the evaluation is completed. Once again, 0.5 to 1.0 mm axial and coronal CT scans are useful and indicated in any patient suspected of an orbital fracture. Globe injury takes precedence over bony repair. If the CT scan reveals bone fragments impinging on the nerve or a hematoma, and if the patient is a surgical candidate, immediate surgical decompression is indicated. If the CT scan does not demonstrate a cause for the altered sight, treat with steroids for five to seven days and evaluate for a response (78). If improvement occurs, continue to follow improvement and delay any repair until vision stabilizes and plateaus. If steroids do not result in improvement during this period, proceed with surgical exploration and repair. The evaluation process will require communication between the neurosurgeon, ophthalmologist and the plastic surgeon in deciding the best treatment option for a patient with altered vision after trauma.

Management

The indications for surgery are diplopia, extraocular muscle entrapment, enophthalmos, and large orbital floor defect (more than half of the floor). Surgery may be performed immediately or postponed until the edema has resolved, usually within a three-week interval from the time of the injury. Exposure of the floor is through a transconjunctival incision, with or without a lateral canthotomy. Subciliary or subtarsal incisions can be used, but have a higher incidence of lid retraction. To date, the best method of reconstruction remains debatable. Many materials are available and used depending on an individual surgeon's preferences. A variety of alloplastic materials have been used including metallic or bioresorbable mesh, and silicone or polyethylene sheets. The primary concern in using alloplastic materials, however, is the potential for infection and extrusion. Our approach is to make a zigzag scalp incision in the temporoparietal area, leaving the pericranium intact. Outline the graft area by incising the pericranium and then harvest "shavings" of the cranium with the pericranium attached by using a wide, thin-bladed, sharp osteotome. The resultant graft takes on a natural curved shape that is ideal for orbital reconstruction.

Complications

See the above discussion on complications in orbitozygomatic fractures.

MAXILLARY FRACTURES

Anatomy

The maxilla occupies the central aspect of the face linking the cranial base above with the occlusal plane of the mandible below. The three vertical supports to the maxilla are the nasomaxillary, the zygomaticomaxillary, and the pterygomaxillary buttresses.

Evaluation

Clinically, a Le Fort fracture presents with malocclusion, maxillary bone instability, bilateral nasal epistaxis, orbital edema, and a step off deformity of the affected bony articulations of the maxilla, to include the zygomaticomaxillary and nasomaxillary buttresses, palpated orally. Clinical evaluation of the dentition and occlusion is important, along with CT scans to accurately evaluate the fracture pattern. A chest radiograph is helpful in evaluating for the



presence of aspirated teeth or tooth fragments. A Le Fort I fracture disconnects the palate and dentition from the mid-face. Le Fort II fractures result in a similar separation, but higher up in the sinus and nasal area. Le Fort III fractures result in a disconnection of the face from the cranium. Disruption of the maxillary arch and palate significantly increases the instability of mid-facial fractures, predisposing the patient to rotation and displacement of the fracture fragments.

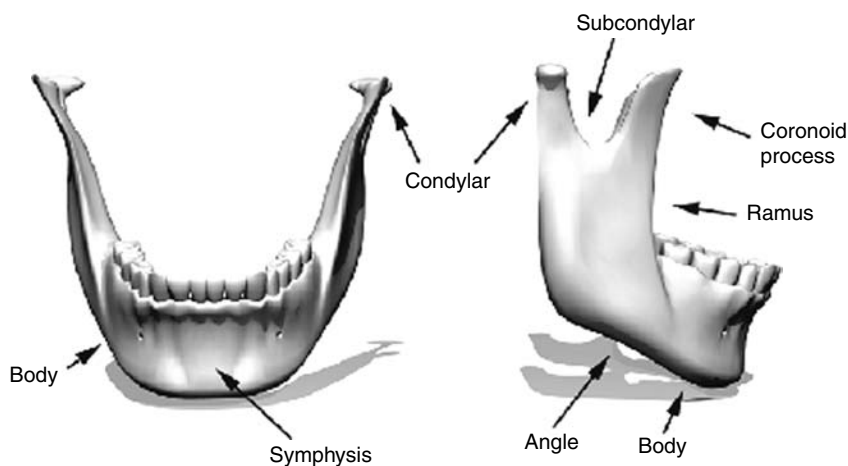
Management

The treatment goals are to restore facial height and projection and reconstruct the preinjury occlusion (79–83). Exposure is through a gingivobuccal sulcus incision, which allows for fracture mobilization, reduction, and rigid fixation. On edentulous patients, the incision is placed on the alveolar ridge due to bone resorption. For adequate fixation, the plate and screws may need to be placed low on the buttress through residual alveolar bone. This may interfere with denture wear. The screws and plates should be removed after fracture healing has occurred. In the interim, the dentures can be relined (84). Placement of arch bars will support the fractured segments by placing the dentition into intermaxillary fixation for the initial fracture reduction, followed by elastics as needed after rigid fixation. Typically, 1.5 to 2.0 mm bone plates are used to fixate the Le Fort fracture along the nasomaxillary and zygomaticomaxillary buttresses. If an intact tooth is avulsed, it should be placed back into the socket and secured carefully using bonding or wire fixation. Comminuted buttresses should be plated with bony continuity. The literature reports the tolerance of 5 mm or smaller gaps in bony continuity, which can be left alone to heal (52). We advocate the grafting of bone into all gaps, especially the vertical and horizontal buttress areas. If bone grafting is required, the fixation plates should be contoured and screwed into place. Second, the bone grafts should be placed into the gaps and secured by screws using the plate holes and/or lag screwed to the bone itself. Placement and contouring of fixation plates is easier to perform prior to bone graft placement.

MANDIBLE FRACTURES

Anatomy

The U shaped mandible has a dentate portion and an articular portion. The dentate mandible has a thick compact inferior border and an alveolar process superiorly. The ascending ramus on both sides ends in a coronoid process and the condylar process. The condyle articulates with the cranium to form the temporomandibular joint. The blood supply of the mandible is the inferior alveolar artery and the surrounding periosteum. To communicate the skeletal relationship between the upper and lower jaw, the Angle classification is used. The Angle classification of dental occlusion refers specifically to the relationship of the mesial buccal cusp of the maxillary first molar to the mesial buccal groove of the mandibular first molar. The occlusion is Class I if the two fit into each other; Class II if the maxillary molar is anterior to



the groove; and Class III if the maxillary molar is posterior to the groove (Fig. 5). The universal numbering system for permanent teeth begins with the patient's right maxillary third molar to the left maxillary third molar (1–16) and then to the left mandibular third molar to the right mandibular third molar (17–32) (Fig. 6).

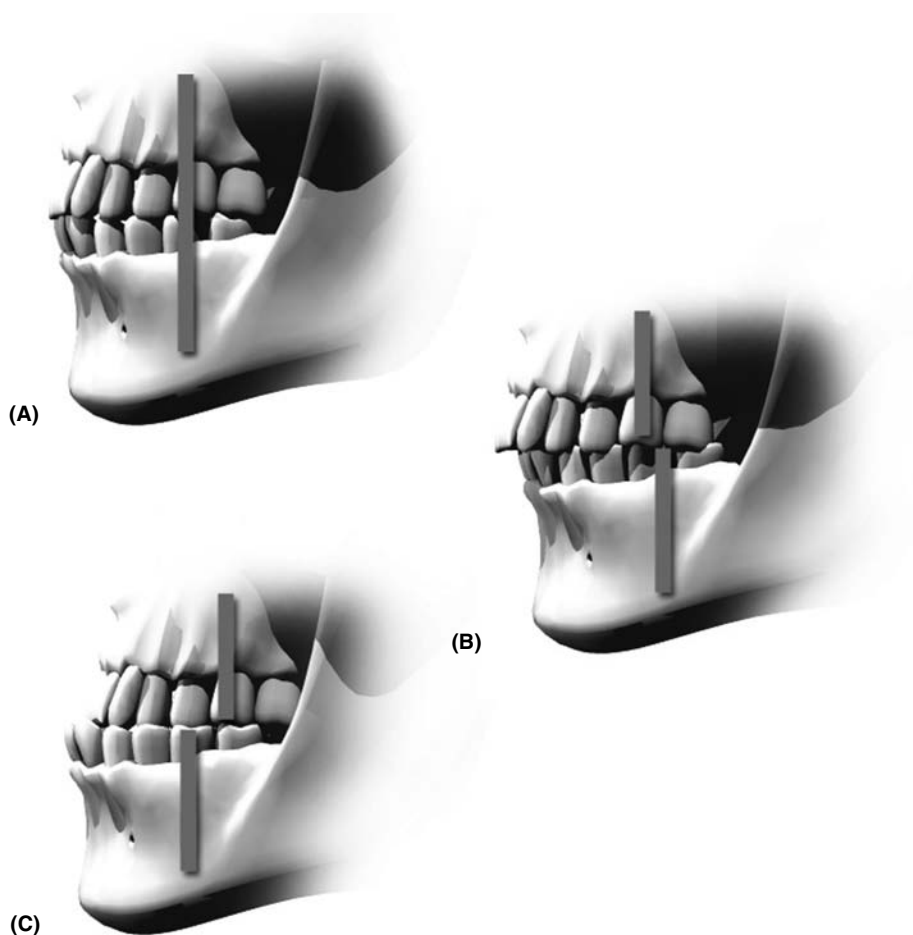


FIGURE 5 Angle's classification of malocclusion: (A) Class I, (B) Class II, and (C) Class III.

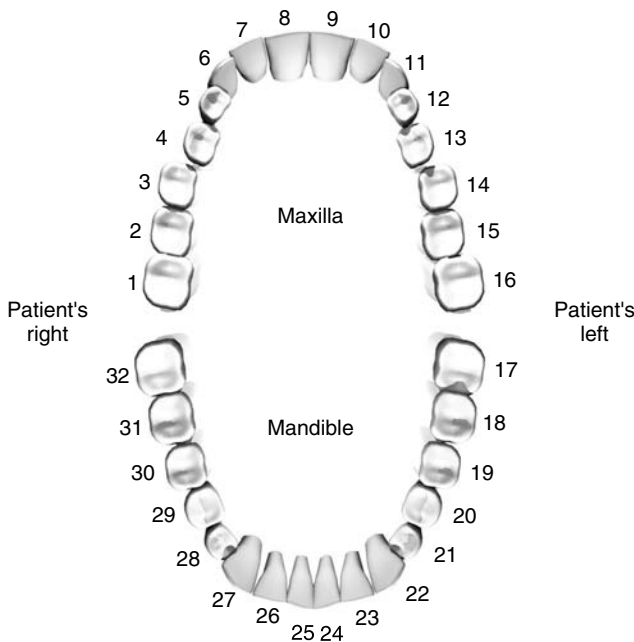


FIGURE 6 Universal tooth numbering system.

Evaluation

Evaluation starts by questioning the patient about any bite alteration. Inspection and palpation of the face along the inferior border and temporomandibular joints may reveal point tenderness and step off deformities. Orally, malocclusion and disruptions of the teeth and dental arch form may be present. Mandibular range of motion may also be disturbed. Anterior traction on the mandible, by gently grasping the mandibular incisors and chin, will often cause pain at the fracture sites. Likewise, gently pushing inward at the angles of the mandible may also reveal fracture sites.

Radiographic evaluation of the mandible is best done with a panorex or CT scan (85–87). Plain films often miss fractures of the condylar processes due to the superimposition of structures. The panorex is most helpful in evaluating the dentition before arch bars are placed. A chest radiograph is helpful in evaluating for the presence of aspirated teeth or tooth fragments.

Management

The goal is the restoration of the lower facial height, chin projection, arch form, and occlusion, which are all important considerations in returning the face to its preinjury condition (88). The location and condition of the fracture dictates the type of reduction appropriate for any given case. Nondisplaced mandible fractures without occlusal disturbances can be treated with a soft diet until healing is complete. Closed reduction is used when a fracture is either nondisplaced or minimally displaced with occlusal disturbances. Arch bars and circumdental wires are applied to the teeth followed by maxillomandibular wires or elastics for two to six weeks to ensure stable reduction. Displaced fractures usually require open reduction with internal fixation using wires or plates. Mandibular fractures should be reduced as soon as possible to minimize pain and soft tissue injury, and reduce the risk of infection. The masseter and temporalis muscles pull upward, and the medial and lateral pterygoid muscles pull inward. Because of the strength of these muscles, displaced fractures often cannot be reduced without the use of paralyzing agents.

BODY AND ANGLE

This fracture site can be reached through an intraoral gingivobuccal sulcus incision with subperiosteal dissection to expose the fracture site. If the fracture is fragmented or other concerns limit an intraoral approach, a submandibular external approach can be used. Non-rigid fixation has a higher risk of complications, such as infection or non-union. Simply placing a wire to hold the reduction is usually inadequate. An impacted tooth in line with the fracture may complicate the reduction. Teeth in the line of fracture should be maintained unless they are split or mobile. Anatomic plate adaptation is important to prevent malocclusion and all efforts should focus on trying to get the best plate adaptation that will closely approximate both the buccal and lingual cortices. Intermaxillary fixation often sufficiently reduces the fracture, so minimal manipulation is necessary before bone plate adaptation and fixation.

Placement of lag screws tangentially is a technique that is easier to conceptualize than apply. The inferior border can be plated with a large mandibular plate and bicortical screws. An arch bar or a plate using monocortical screws can be used to overcome tension and secure the reduction. Posteriorly, the fracture fixation can be accomplished intraorally via a right angle drill or through a percutaneous trocar.

SYMPHYSIS

This area is usually handled with open reduction and fixation with a mandibular fracture plate on the inferior border and an arch bar on the superior aspect. Exposure of this area with a gingivobuccal sulcus incision focuses on minimizing trauma to the mental nerves. After fixation, reattaching the mentalis to the bone with a suture to the periosteum or to a screw will help prevent chin ptosis.

RAMUS, CORONOID, AND CONDYLE

Fractures of the ramus and coronoid are rarely significantly displaced and can be managed by closed reduction/intermaxillary fixation. The intracapsular fracture of the condyle is a specific fracture with a high risk of ankylosis. If there is no malocclusion, the patient can be placed on a soft diet. Jaw mobility should be encouraged and checked frequently. If malocclusion is present, the patient should be placed into maxillomandibular fixation that is released in 10 to 14 days. The patient is then placed into guiding elastics. Open reduction of condylar and subcondylar fractures are debatable but should be considered when the following conditions are present (89–94):

- Displacement of the proximal segment of 30° or more from the axis of the ascending ramus
- Shortening of the ascending ramus with telescoping of the fragments and open bite
- Dislocation of the condylar head from the temporomandibular fossa
- Inability to obtain adequate occlusion with closed reduction

In addition to these, the concomitant presence of a mid-facial fracture is an indication for the open treatment of mandibular condyle fractures. Bilateral subcondylar fractures associated with mid-facial fractures are managed with open reduction and internal fixation of the condylar neck, at least unilaterally, to reestablish the vertical dimension of the face.

Submandibular, preauricular, and intraoral ramus incisions to expose the condylar process are all valid approaches. Endoscopically assisted open reduction of condyle fractures is a helpful technique to avoid an unsightly neck incision (95).

MAXILLOMANDIBULAR FIXATION

When applying arch bars, attention to the gingiva is important. The wires around the teeth should be placed without crushing the gingiva, which can cause edema, necrosis, and subsequent aesthetic concerns. If possible, circumdental wires should be avoided around the central incisors. In appropriate patients, the use of bonded brackets to manage fractures should

be considered. The bonding of brackets to the teeth by an orthodontist will save operating time, and the result is much more tolerable by the patient (96–98).

Interdental wiring or elastics are potentially hazardous in patients with an altered mental status. Wire cutters or scissors should be kept at the bedside to be used to release the fixation if vomiting or airway problems develop. Patients who are discharged to their home should be instructed in removing their intermaxillary fixation if problems develop.

TEETH IN THE LINE OF A FRACTURE

Teeth are retained whenever possible (99–101). Teeth that are completely avulsed should be re-implanted as quickly as possible after careful repositioning of the alveolar fragment and stabilization with an arch bar. If the alveolar fracture is part of a fracture extending to the inferior border, the avulsed tooth is not re-implanted. Similarly, a tooth whose root is transected or split by the fracture should be removed. A fracture of the crown with a stable root should be saved and considered for dental restoration. If the entire crown is fractured it should be left in place to be addressed by a dentist at a later date.

FRACTURES IN CHILDREN

In childhood, the mandible is weakened by numerous unerupted and developing permanent teeth, which limit the amount of bone and create regions susceptible to fracture. The treatment of children with mandibular fractures is different from that of adults because of the presence of deciduous teeth, unerupted teeth, and tooth buds. Most fractures are best managed by closed reduction. Between ages two and six, a variable number of deciduous teeth will allow for placement of arch bars. Between 9 and 12 years of age, most children have a mixed dentition, with multiple loose or missing teeth, making arch bar placement difficult. In these cases, circummandibular wires and piriform rim wires may be required for fixation. Most fractures in children are immobilized for 7 to 14 days and the patient is allowed to return to function 10 to 21 days after surgery (102).

Condylar fractures in children are often intracapsular with a high risk of ankylosis and/or growth disturbances. The condyle is the growth center for the mandible. Brief maxillomandibular fixation (five to eight days) followed by vigorous postoperative mobilization is recommended for these fractures.

EDENTULOUS PATIENT

Open reduction and internal fixation, with the use of dentures to assist in the initial reduction, is the recommended approach. The use of a large reconstruction fixation plate is recommended (103). The atrophied bone and poor blood supply make these fractures particularly difficult (104). Nonunion is the most feared complication when treating these fractures. Contributing to the difficulty in treating these fractures is the lack of thick bone to place screws and the lack of teeth for intermaxillary fixation. Primary bone grafting is a consideration in the treatment of body fractures in the atrophied mandible.

POSTOPERATIVE CARE

The primary concern in patients with mandibular fractures treated with maxillomandibular fixation is the airway. The endotracheal tube is left in place until the patient is awake and able to maintain their airway. The prophylactic use of antibiotics has been widely accepted. Nutrition, consisting of protein and complex carbohydrates, is important. Lastly, oral hygiene needs to be emphasized. Brushing and rinsing the mouth with chlorhexidine (Peridex) mouth rinse or half-strength hydrogen peroxide is recommended.

COMPLICATIONS

The most common complication of compound mandibular fractures is infection. Therefore, pre and postoperative antibiotic coverage is recommended (105,106). In cases in which the infection is confined to the soft tissues, incision and drainage will suffice. If a carious tooth causes infection, antibiotics and tooth removal are indicated. Patients should be maintained in rigid fixation until the fractures are healed (usually four to six weeks). It has been shown that fracture segments will heal within a region of infection if they are rigidly fixed. If the infection involves bone fragments, aggressive debridement of bone, soft tissue, teeth, and loose hardware are indicated. An extraoral approach is recommended. All devitalized bone is removed until bleeding is visualized. This is followed by accurate anatomic reduction and fixation with large reconstructive plates. Immediate or delayed bone grafting follows, if appropriate (107). An alternative approach is to debride the area and maintain external fixation followed by delayed reconstruction in three to four months.

Nonunion is a fracture that has not healed within six months. Mobility and infection are the most common cause of nonunion. The area of nonunion should be explored and intervening soft tissue and necrotic bone removed until healthy bleeding bone is visualized. Similarly an extraoral approach is recommended. The bony segments should be aligned and rigidly fixated with a large reconstruction plate and bone grafted, as needed.

Malunions, and as a result malocclusions, are rare. If minimal, orthodontia can be considered. If the problem persists, a full orthognathic evaluation, with planned surgical osteotomies, may be necessary to restore preinjury occlusion.

Injuries involving the inferior alveolar and the mental nerves are not unusual. Most injuries are due to stretching or compression and will spontaneously resolve over many months. If the problem persists, a CT scan and panorex can be obtained to rule out any mechanical cause that potentially can be treated.

COMPLEX FRACTURES

The two basic sequences to pan-facial fracture repair have traditionally been from bottom to top or from top to bottom (108,109). The bottom to top technique is based on the fact that the mandible can be reconstructed to provide an intact relationship for positioning of the maxilla. Positioning of the maxilla relies on the proper seating of the condyle in the glenoid fossa. In the top to bottom method, mid-face reconstruction can precede the fixation of the mandible if adequate bony keys are available to ensure proper maxillary positioning. Reconstruction of the outer facial frame is the key to successful reconstruction.

First, reconstruct the outer facial frame to include the zygomatic arch, zygoma, and frontal areas. Second, reconstruct the inner facial frame to include the nasooethmoid complex, zygomaticofrontal, and infraorbital rim. Third, reconstruct the maxilla at the Le Fort level by plating the buttresses. Lastly, temporary IMF followed by open reduction and internal fixation of the mandible is done. The advantage of the top to bottom sequence is it allows for the closed treatment of subcondylar fractures.

A specific complex injury pattern is the Le Fort fracture with a midline fracture of the palate, and a mandible fractured through the body and at both condyles. With the loss of the dental arch form in both jaws, the resultant facial reconstruction is often too flat and wide. The key to reconstructing this facial fracture pattern is to first open and plate both condylar processes. Next, the mandibular body fracture can be plated with the width of the mandibular arch being controlled by the condyles seated firmly in the glenoid fossae. The rest of the reconstruction can proceed from bottom to top after the split maxillary segments are joined to the intact mandible with arch bars, and plated.

Many facial fractures are not isolated but involve a combination of the fracture patterns that have been described. It is important that the approach be well planned and organized. Any facial bone fracture must be aligned by addressing some or all fracture sites with adjacent bones. The need for exposure of all the buttresses of a fractured bone is governed by the difficulty of reduction and by the need for stabilization, both of which relate to the displacement and comminution of the fracture. When multiple areas of the face are fractured, an order of

treatment needs to be developed. The exact order of treatment is not as important as the development of a plan that permits flexibility and accurate positioning of the fractured areas. The operative procedure for complex fractures can be lengthy and requires the utmost attention to efficiency, with appropriate attention to critical surgical steps. If occlusal fractures are present, the placement of arch bars and maxillomandibular fixation is a reasonable first step. This can be followed by the exposure of all affected areas.

Since grafting orbital defects to reduce volumetric disturbances is difficult in a field of massive edema, I personally leave the orbital areas free from dissection until I am ready to address the condition. In patients with complex orbital fractures, grafting and anatomic correction may be difficult and "tight" due to traumatic edema, but yet is necessary to obtain the best results. The results from the initial surgery will set the stage for further reconstruction. Secondary orbital correction is difficult and therefore, the degree of correction during the initial surgery is paramount to the overall outcome.

CONCLUSION

The management of facial fractures ranges from the simple to the highly complex. The difficulty arises in the appropriate planning and, secondarily, in the execution during the definitive repair. The best preparation starts with the initial evaluation and diagnosis. During the preoperative planning, flexibility and adaptation to surgical findings are critical. Before surgery, it is critical to communicate to the patient and family the operative plan and the possibility of deviation depending on operative findings. The patient and family need to be aware of the severity of the injury and the high likelihood that further surgeries and treatment will be needed.

The operative procedure itself should focus on making sure that appropriate incisions to adequately reduce and fixate the fractures are utilized. It is helpful to review the contours of the facial skeleton before surgery. Avoidance of common errors that lead to widened faces, loss of projection, or flattening of contours is critical. It is important to address the soft tissues appropriately. All lacerations and incisions should be closed in layers with emphasis on re-suspending the soft tissues to prevent ptosis. Additionally, develop a lower eyelid approach to minimize the possibility of lower lid retraction. Lastly, all patients require follow up and education in regard to potential complications. The use of consultants and adequate assistance are all part of a successful team approach to the management of facial fractures.

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20 The Management of Established Post-Traumatic Craniofacial Deformities

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BACKGROUND

Over the past generation, the principle of precise open reduction and rigid internal fixation with the use of low profile, biocompatible hardware has revolutionized the acute management of facial trauma. The ability to achieve stepwise skeletal fragment reassembly, and accurate reestablishment of facial projection, contour, and buttress support, has become more consistent due to the availability of rigid internal fixation devices. Concurrently, modern techniques of craniofacial reconstruction developed by Tessier and others have been extended for use in trauma reconstruction. With this has come new routes to the injured craniofacial skeleton (e.g., coronal and transconjunctival incisions, multi-incision minimal access approaches, endoscopic techniques), the routine use of split calvarial bone grafts, and the introduction of customized instrumentation (specialized power saws and drills, retractors, periosteal elevators, lighting, etc.).

However, despite the advances made in acute reconstruction, clinically significant post-traumatic deformities are still routinely encountered following primary repair. Issues that may compromise a surgeon's ability to anatomically restore a patient at the time of acute injury include: associated massive soft tissue destruction (coverage and/or lining); extensive bony comminution; unstable medical condition preventing a patient from undergoing acute reconstruction; and suboptimal surgical technique.

While applying many of the same concepts of care, the evaluation, planning, and modalities of treatment for established post-traumatic craniofacial deformities can differ from that for acute injuries. It is often necessary for the surgeon to see through the mask of distortion and to envision a three-dimensional unscrambling of the secondary deformity in order to understand how to properly reestablish anatomic normalcy. Soft tissue loss and contracture, bony deficit, resorption, and remodeling can confound assessment. Comparison to unaffected contralateral structures is helpful but not always an option.

GENERAL PRINCIPLES

Ultimately, proficient treatment of a complex secondary craniofacial deformity requires that the surgeon follow some general principles.

Preoperative Assessment

1. Obtain as much information as possible regarding the original injury and prior reconstructive efforts
2. Determine the patient's perception of their own deformity/dysfunction
3. Perform meticulous clinical examination paying attention to symmetry, contour, projection, texture, vascular perfusion, and function

4. Identify any disturbances in airway, vision, cutaneous sensation, facial nerve function, temporomandibular range of motion, or dental occlusion
5. Consider the integrity of all tissue layers: lining, support, and coverage
6. Utilize modern imaging techniques to carefully assess skeletal deficit and/or displacement
7. Carefully explain options, prospects, and limitations of surgery; elicit patient's concerns and wishes regarding possible reconstruction
8. Perform careful preoperative photography and obtain dental models (when applicable) for the purposes of evaluation, planning, and documentation
9. Make patient aware of the possible need for staged reconstruction

Surgical Treatment

1. Refracture and anatomically reduce dislodged bony structures when possible
2. Replace missing parts with like autologous tissue—vascularized and local tissue when possible; proceed stepwise up the reconstructive ladder
3. Utilize camouflage onlay techniques only for minor deformities
4. Employ minimal access techniques when appropriate

OPERATIVE TECHNIQUES

Classic fracture configurations are not always easily identifiable and any unique combination of craniofacial injury pattern may be discovered. However, for the purposes of simplification, reconstructive solutions to established post-traumatic facial deformities will be presented here as they may be applied to different anatomic regions.

Cranial Defect

Secondary cranial deformities vary from minor depressed fractures to major segmental full-thickness defects. Smaller contour deformities that underlie regions of hair-bearing scalp typically do not require correction. Those in the forehead region, however, might represent an important aesthetic irregularity and can be addressed using any of a variety of either autograft (preferable—e.g., split cranial bone chips, fat grafts), homograft (e.g., acellular dermal matrix, demineralized bone), or alloplastic (hydroxyapatite cement paste, porous polyethylene, etc.) onlay materials. These procedures can occasionally be performed through limited, local scalp incisions for smaller defects (endoscopic assistance may be considered when using a hair-bearing incision to correct a lower forehead deformity), or through coronal incisions when greater exposure is required.

Reconstruction of a full-thickness cranial defect is usually a more complex intervention necessitating neurosurgical involvement to separate the overlying coronal flap from the dura. Following the principles outlined above, it is preferable to replace missing tissue with like tissue. Smaller defects can be repaired using split cranial grafts harvested in situ from adjacent cranium. For larger defects, full-thickness cranial grafts are required and can be harvested with the help of a neurosurgeon. A template of the recipient site can be obtained and used to trace a replica over an appropriate donor location (often the contralateral cranium) so as to avoid the harvest of surplus bone and to expedite the tailoring of the graft to the defect.

In certain cases of extremely large full-thickness cranial defects, it may be determined that split calvarial bone grafting from the residual cranium is contraindicated or unfeasible. Alloplastic biomaterials such as methylmethacrylate or porous polyethylene may be utilized to advantage in those instances. Three-dimensional computed tomography (CT) scans can be used preoperatively to allow for the fabrication of synthetic implants that closely match the defect.

Case 1

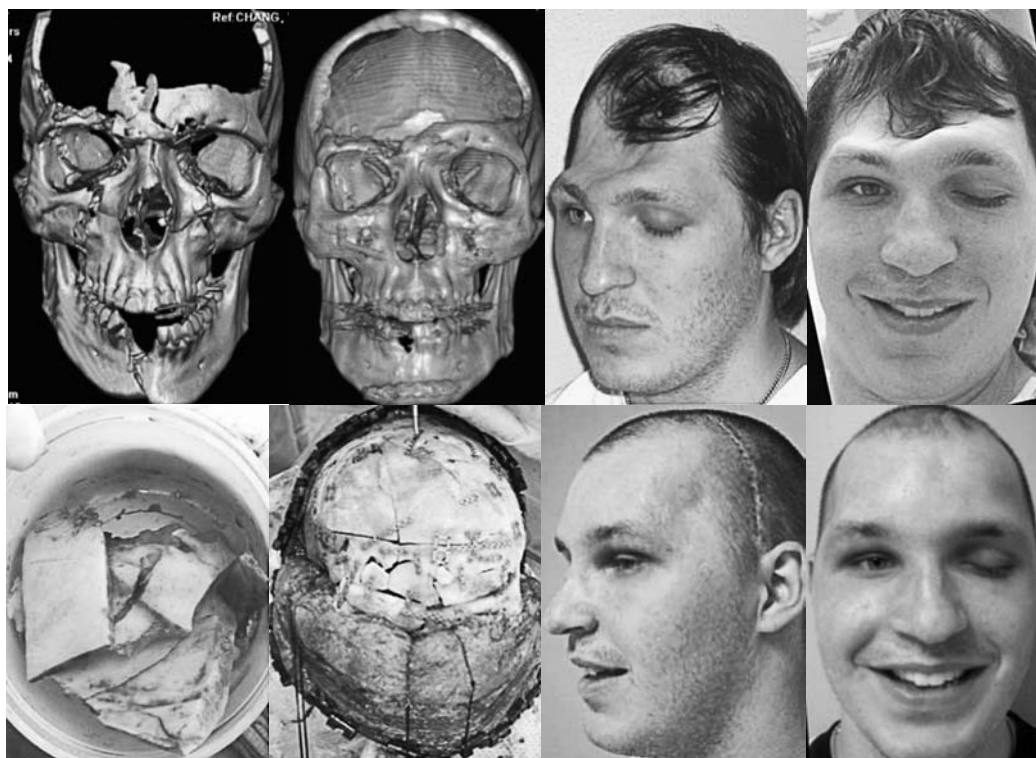
At 2 years, 10 months of age, the following patient fell from a second story window suffering a right frontoparietal compound fracture, subdural hematoma, and cerebral contusion. Neurosurgery discarded approximately half of the comminuted right parietal bone due to frank contamination, and four months later repaired the defect with a titanium mesh. At four years of age, he presented with an exposed, fractured mesh that was removed. At four years, five months of age, he underwent a definitive cranioplasty using split cranial bone harvested from the contralateral parietal bone. Resorbable fixation was used. Lower right CT scan is from 11 months postoperatively. This has remained stable now for five years.



Cranial Defect: Case 1.

Case 2

An 18-year-old boy riding a motorized scooter unhelmeted, collided with a utility pole at 40 mph. He suffered panfacial fractures and comminuted frontal fractures, involving cerebral tissue herniating through his forehead laceration. Decompressive craniectomy was performed emergently. Open reduction and internal fixation of panfacial fractures and frontal sinus cranialization were performed on day 12 post-injury. Cranioplasty at 11 months post-injury required the use of split cranial bone to reconstruct the right supraorbital ridge and lower forehead regions, which were discarded at the time of the initial craniectomy. The remainder of the frontal bone was reassembled from stored bone fragments using titanium fixation. Patient is stable at four months following cranial reconstruction.



Cranial Defect: Case 2.

Frontal Sinus

A minimally displaced ($<1\text{--}2\text{ mm}$) isolated anterior table frontal sinus fracture usually does not result in the development of a clinically important deformity. The magnitude of a more significant outer table depression can sometimes be obscured by early post-injury edema. The early concealment of the concavity can lead to patient reluctance to provide consent to a primary reconstructive procedure. Unfortunately, this may also result in the later presentation of an established forehead deformity. A surgeon's options when confronted with a secondary frontal sinus outer table concavity include the onlay camouflage techniques discussed above for small contour deformities of the cranium. Alternatively, refracture and reposition of a frontal sinus outer table depression is also a suitable option since it does not necessitate an intracranial procedure. Using a narrow side-cutting burr, the fracture edges can be retraced and the mobilized bony fragment gently levered into anatomic reduction.

For more extensive frontal sinus fractures involving the frontonasal ducts, a low threshold for sinus obliteration should be maintained at the time of primary management. Outflow obstruction of an intact frontal sinus can lead to sinusitis or mucocele (or mucopyocele) formation that over time can result in significant adjacent bone resorption and meningitis. Management of a frontal sinus mucocele involves meticulous and complete sinus exenteration (including careful burring of all bony surfaces of the sinus) and duct obliteration. Associated skeletal deformities should be repaired at the same time using standard craniofacial techniques.

Case 3

A 40-year-old patient presented 13 years following facial fractures sustained in a tire explosion accident. Reconstruction and obliteration of frontal sinus performed elsewhere. At the time of presentation, the patient reported a two-year history of a growing mass in the left medial canthal

region. Preoperative CT scan revealed a 25×17 mm mass extending from the left frontal sinus remnant into the left orbital space, obstructing the visual field, consistent with a mucocele. The sinus was accessed via a window osteotomy performed with a 1.2-mm side-cutting burr to limit kerf, which required extension in order to allow adequate access for meticulous mucosal excision, burring of the sinus cavity, and duct obliteration. Split cranial bone grafts were used to reconstruct the medial and superior orbital walls. Diploic bone and a pericranial flap were used to obliterate the duct. Patient is stable at two years post-reconstruction.



Frontal Sinus: Case 3.

Case 4

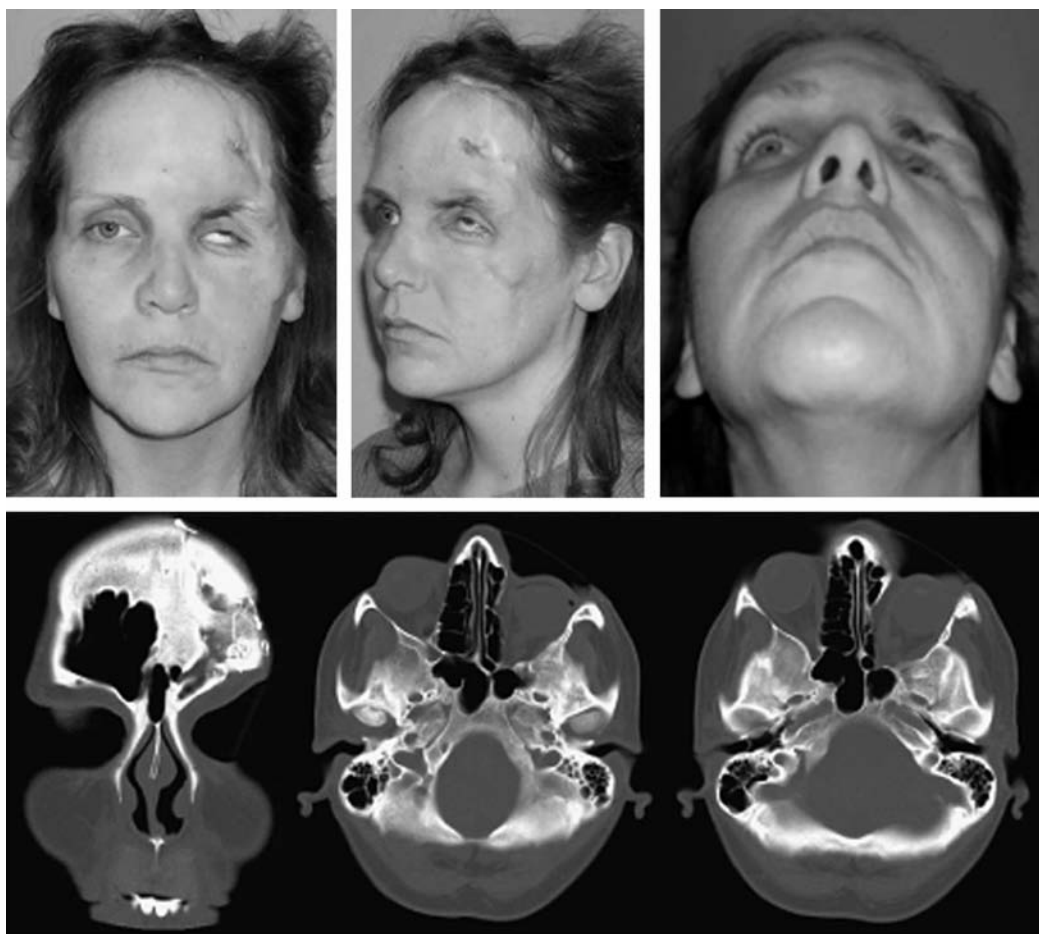
A 20-year-old patient suffered a depressed frontal sinus (outer table) and nasal fractures playing rugby. A 1.2-mm side-cutting burr was used to refracture the displaced segment in order to allow it to be levered back into anatomic position with minimal bony kerf created. Patient shown at three weeks post-reconstruction and has remained stable for five years.



Frontal Sinus: Case 4.

Case 5

A 36-year-old patient struck by a motor vehicle as a pedestrian 20 years ago. She underwent multiple reconstructive procedures elsewhere in the initial five years following the injury. She presented with a several month history of a left forehead fistula expressing purulent material, with exposed hardware and bone. Her left eye has minimal vision and she routinely wears an eye patch for cosmesis. Her CT scan demonstrated an intact right frontal sinus with communication to the residual left frontal sinus and reconstructed left frontoorbital region. First stage of treatment included a left frontal craniectomy to debride approximately a 10 cm² area of osteomyelitic bone, a meticulous frontal sinus exenteration and obliteration, and a primary excision and repair of the fistula site. Subsequent reconstruction must await an appropriate course of intravenous antibiotics and confirmation that the infection has cleared. Thereafter, the basic principles of craniofacial reconstruction will apply. This case highlights the importance of proper primary treatment of the frontal sinus, and the need to control infection prior to pursuing secondary reconstruction.



Frontal Sinus: Case 5.

Nasoorbitoethmoidal Injuries (Traumatic Telecanthus)

In addition to marked nasal deformity and disruption of the lacrimal drainage system, nasoorbitoethmoidal (NOE) injuries may result in traumatic telecanthus. The latter deformity is avoided at the time of primary reconstruction by properly reducing the canthal-bearing fragment of bone. In cases with extensive comminution, the key fragment may be too small to

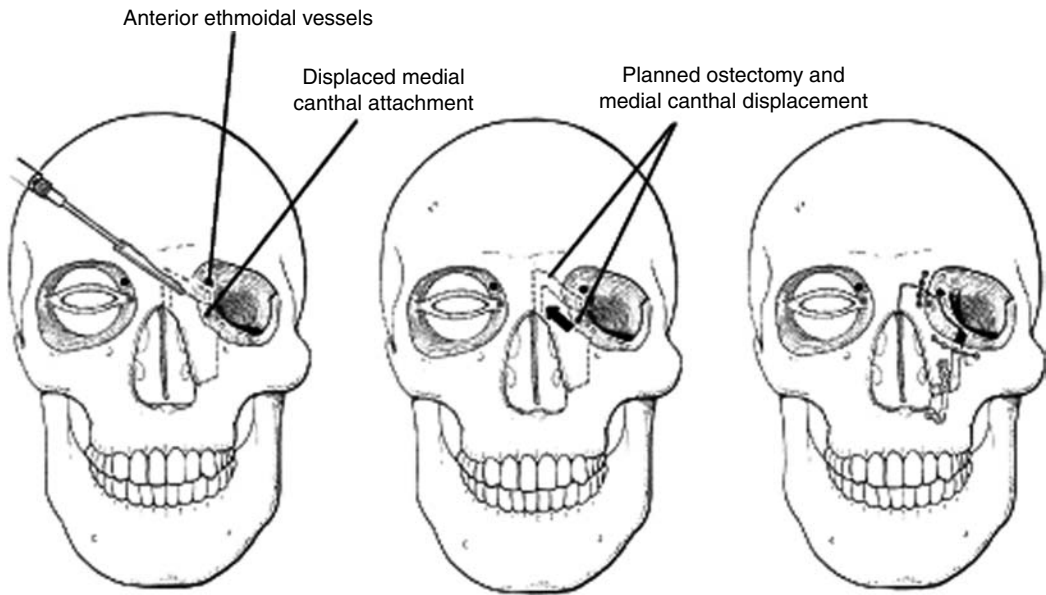


FIGURE 1 Medial orbital osteotomy and displacement for the correction of medial canthal malposition. *Source:* Adapted from Ref. 2.

fixate and thus a precise reduction of the medial canthal tendon must be achieved using a transnasal medial canthoplasty.

Adhering to the general principles outlined above, the most effective approach to correction of an established post-traumatic telecanthus involves refracture and anatomic reduction of the canthal-bearing fragment. This requires osteotomies peripheral to the lacrimal fossa and caudad to the anterior ethmoidal vessels, as well as a parasagittal osteotomy allowing for medial displacement of the fragment (Fig. 1). By avoiding the use of a transnasal medial canthoplasty, the medial orbital osteotomy and displacement allows one to circumvent the need for tendon detachment and the challenging task of securely fastening it in a more anatomic position.

Case 6

A 13-year-old patient was kicked in the face by a horse. This resulted in a left-to-right naso-orbital fracture displacement and associated left enophthalmos and right telecanthus with characteristic canthal rounding. An initial left orbital floor repair eight months earlier utilizing a silastic implant was associated with a suboptimal outcome. A refracture and leftward reposition of the composite bilateral naso-orbital complex was performed. *Source:* From Ref. 11; photos courtesy of Dr. Henry K. Kawamoto, Jr., DDS, MD, University of California at Los Angeles.



Nasoorbitoethmoidal Injuries: Case 6.

Case 7

A 27-year-old patient was struck by a car while riding his bicycle, suffering frontal and NOE fractures. An associated dural tear was repaired initially, but definitive frontal and NOE repair

was delayed. His established deformity included bilateral telecanthus with anti-Mongoloid slanting of the palpebral fissures. This was addressed by reversing the skeletal displacement, performing bilateral medial orbital osteotomies, and repositioning the canthal-bearing fragments in a cephalomedial direction. *Source:* From Ref. 11; photos courtesy of Dr. Henry K. Kawamoto, Jr., DDS, MD, University of California at Los Angeles.



Nasoorbitoethmoidal Injuries: Case 7.

Case 8

A 31-year-old patient was involved in a motor vehicle collision resulting in a right Le Fort I, NOE, and zygomaticomaxillary complex fractures (ZMC). He presented with established deformities including right telecanthus, vertical dystopia, midface shortening, malar flattening, and a saddlenose. His telecanthus was corrected by repositioning the right canthal-bearing medial orbital complex in a cephalomedial direction. *Source:* From Ref. 11; photos courtesy of Dr. Henry K. Kawamoto, Jr., DDS, MD, University of California at Los Angeles.



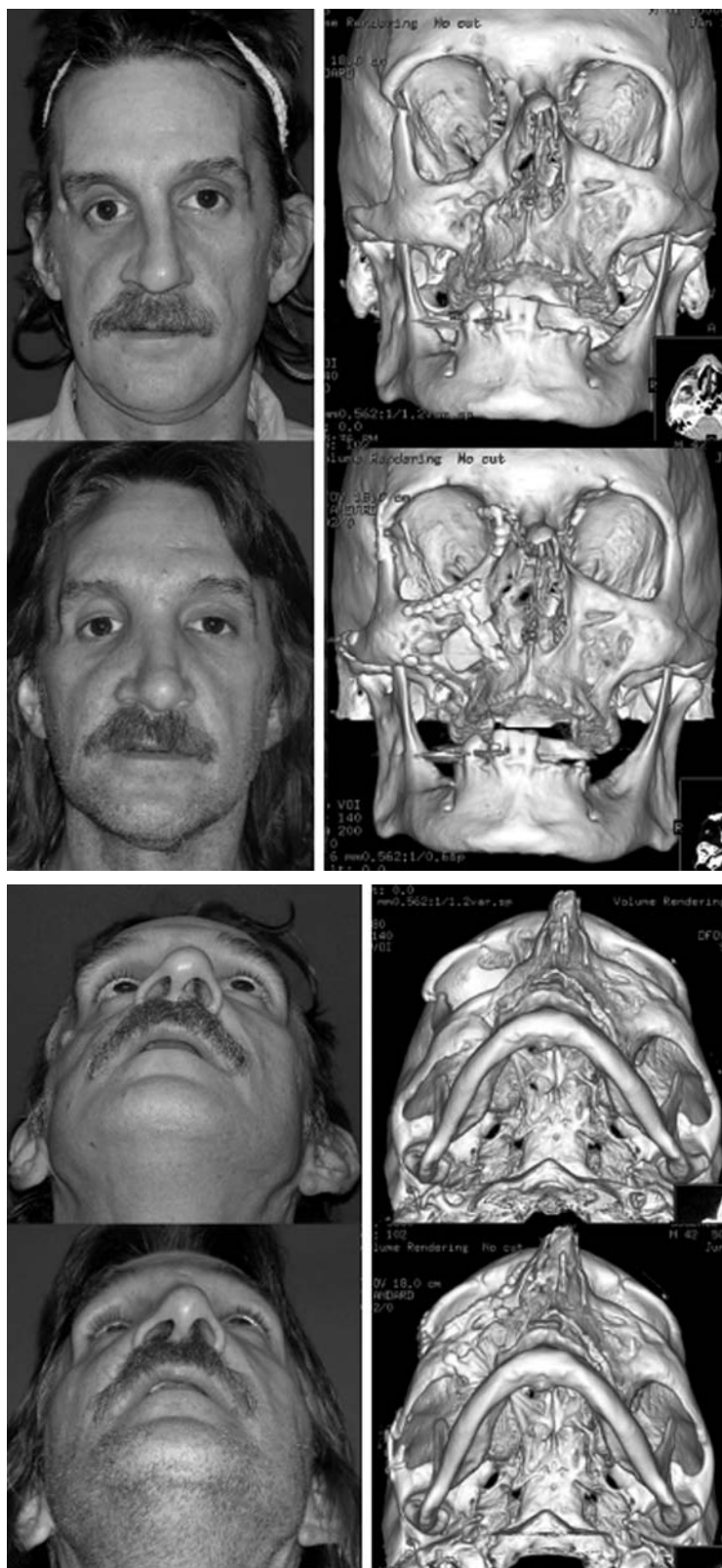
Nasoorbitoethmoidal Injuries: Case 8.

Orbito-Zygomaticomaxillary Deformity

Enophthalmos is perhaps the most commonly encountered secondary post-traumatic deformity. It may arise from either an isolated orbital injury or a more involved orbito-zygomaticomaxillary trauma. In correcting isolated orbital defects, all four bony walls should be evaluated carefully and their integrity reconstituted, preferably with split cranial bone grafts. More complex fractures are best treated by refracture and reposition, when feasible. In cases in which the zygomaticomaxillary region is deficient in bone stock, and depending on the particular patient circumstances, extensive onlay bone graft reconstruction may be a practical and effective mode of anatomic restoration.

Case 9

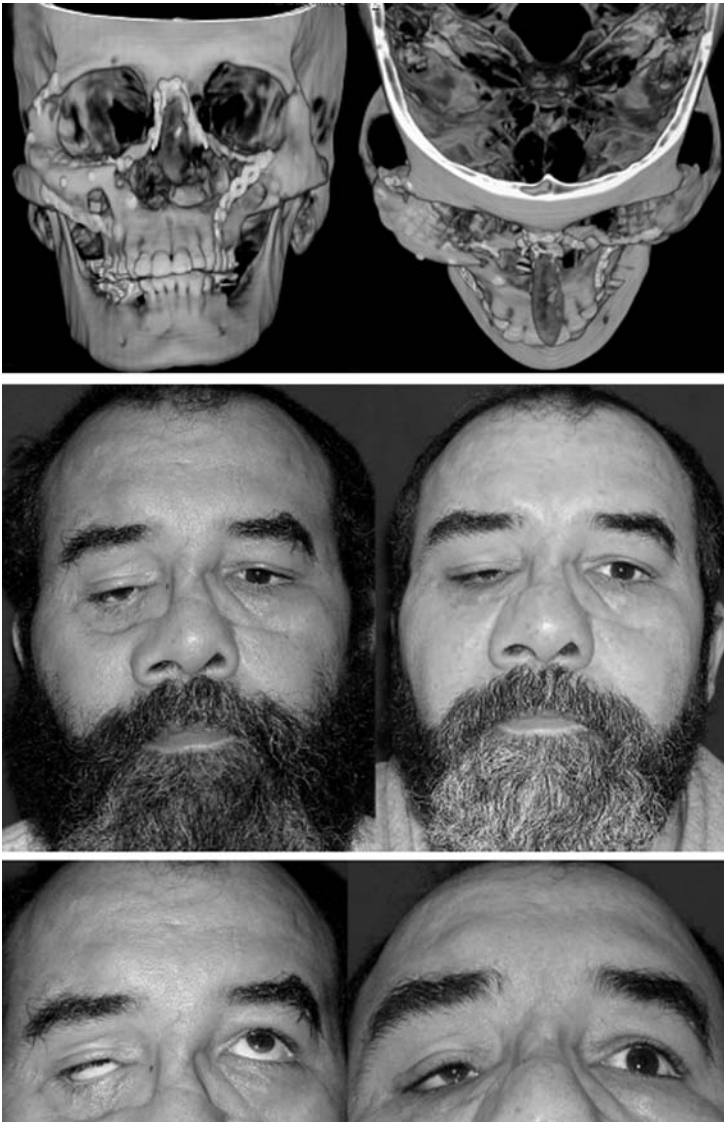
A 41-year-old patient involved in a motor vehicle accident with right nasoorbital and zygomaticomaxillary injuries is shown. Associated thoracic injuries prevented early repair. At five and half months post-injury, he underwent a right ZMC fracture reposition involving a coronal, lateral upper blepharoplasty, lower lid transconjunctival, and right upper buccal sulcus incisions. An open septorhinoplasty was later performed at a second stage (following the postoperative CT shown) that involved osteotomies with outfracture of the right and infracture of the left nasomaxillary bones, and septal centralization maneuvers. Despite improvement in malar depression and vertical orbital dystopia, some signs of residual enophthalmos remain. The patient is satisfied with the correction and has refused further intervention.



Orbito Zygomatocomaxillary Deformity: Case 9.

Case 10

A CT scan of 43-year-old man is shown, demonstrating established craniofacial deformities sustained in an industrial accident seven years prior to presentation and following multiple surgical interventions elsewhere. Note on CT that the right ZMC is displaced/rotated in an antero-caudad and lateral direction, with an associated increase in right orbital volume. Note the reduction in midface bone stock, particularly the bilateral medial buttresses, right lateral buttress, and right inferior orbital rim. Note the presence of large orbital floor reconstruction meshes bilaterally. Postoperative photos are from 21 months following onlay split calvarial bone grafts to the right orbital floor, right inferior orbital rim, and right nasal sidewall; soft tissue resuspension of right midcheek; and right lateral canthoplasty. Surgical access was via prior lid-cheek scar and lateral upper blepharoplasty incisions. An 8-cm right parietal incision enabled harvest of bone grafts material. The patient was pleased with the gains achieved from this procedure and has refused further intervention to address residual deformities.



Orbito Zygomaticomaxillary Deformity: Case 10.

Case 11

A 38-year-old patient was an unhelmeted snowmobile driver who collided with a stone wall nine months prior to presentation. Overriding medical concerns prevented primary repair of facial fractures. A right zygomaticomaxillary fracture reposition was performed via upper buccal sulcus, lateral upper blepharoplasty, and lower lid transconjunctival (without canthotomy) incisions. Five-month postoperative photo is shown below.



Orbito Zygomaticomaxillary Deformity: Case 11.

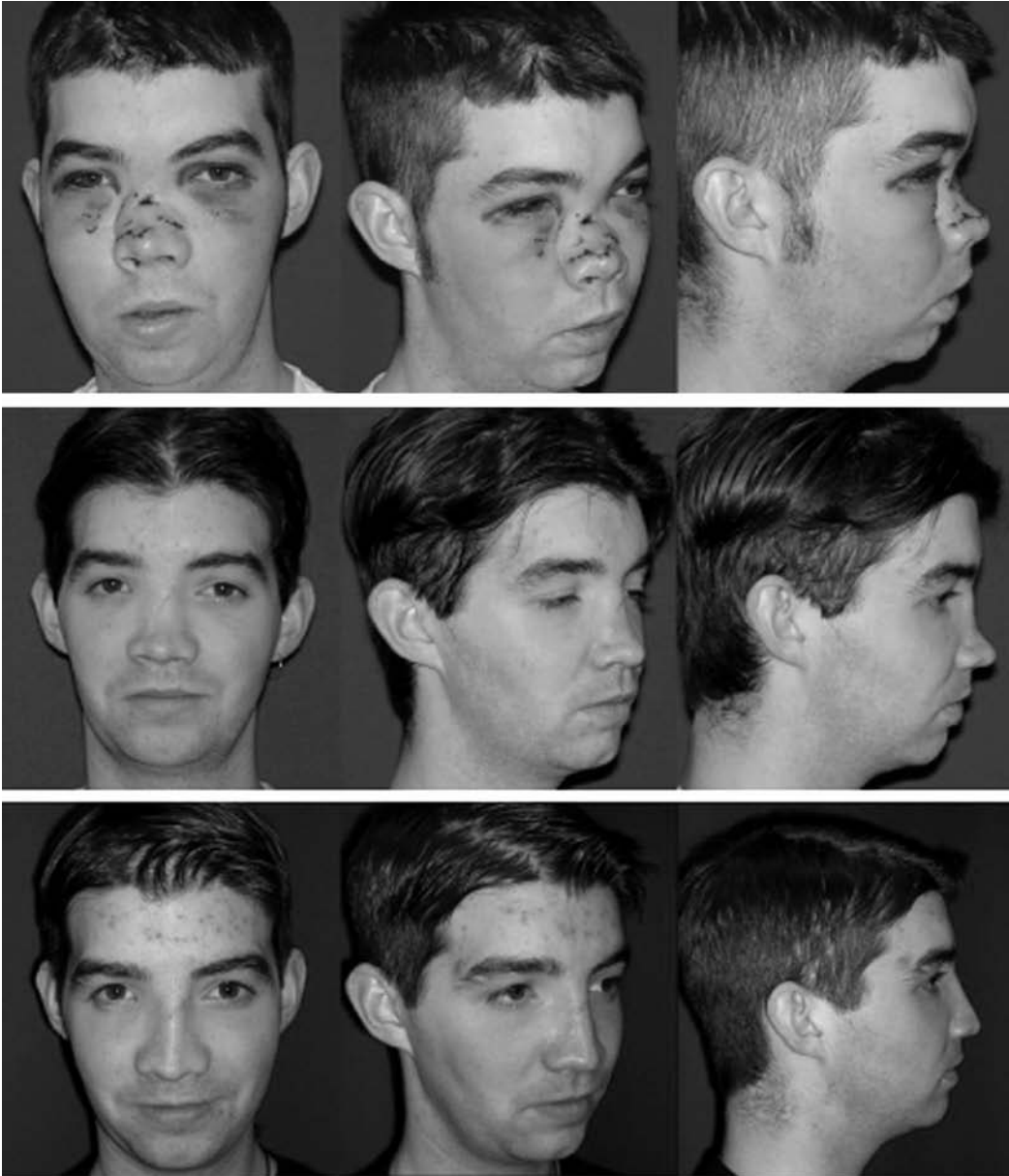
Nasal Injury

For established post-traumatic nasal deformities involving lateral deviation and/or collapse of the bony pyramid, standard septorhinoplasty techniques apply well. The use of lateral, medial, and transverse osteotomies, as well as greenstick fracture of the perpendicular plate of the ethmoid may be required to properly mobilize and reduce the displaced bony fragments. Submucous resection, cartilage scoring, bony spur excision, caudal septal repositioning, suture modification, and batten strut grafting of the septum are all valuable maneuvers to consider in attempting to correct a marked septal deviation. In the case of major NOE injuries, major impaction and comminution of the nasal osseocartilaginous skeleton may occur. Primary bone grafting is indicated if nasal projection is deficient after open reduction and fixation is completed. The correction of an established saddlenose deformity requires a supporting bone or cartilage graft. A cantilever graft can be inserted through a closed rhinoplasty approach or through a preexisting scar in the nasion region when available. The base of the graft should be contoured and beveled in order to maximize osseous contact with the underlying nasofrontal region, and to achieve the desired angle of projection. Two titanium microscrews lagged through the graft will usually suffice in providing stable fixation (and these can be placed percutaneously through a stab incision when the graft is inserted through a closed technique). The source of the bone graft is debatable, and depends on the reconstructive needs as well as the patient age (which impacts graft quality at different locations). Certainly, split cranial bone is the most convenient and well-tolerated donor site, and can provide easily contoured bone that will maintain its volume over the long term.

Case 12

A 23-year-old patient was involved in a motor vehicle accident leading to bilateral NOE and maxillary fractures. Primary open reduction internal fixation, orbital floor and medial wall bone grafting, and right transnasal canthoplasty were performed but resulted in a saddlenose deformity. Secondary reconstruction at 10 months post-injury involved a split calvarial cantilever bone graft measuring approximately 5 cm in length. Surgical access for bone graft placement was via a preexisting nasal dorsal scar, which was revised at the same time.

The patient has persistent mild right telecanthus but has refused further corrective intervention. Two-month postnasal reconstruction is shown below.



Nasal Injury: Case 12.

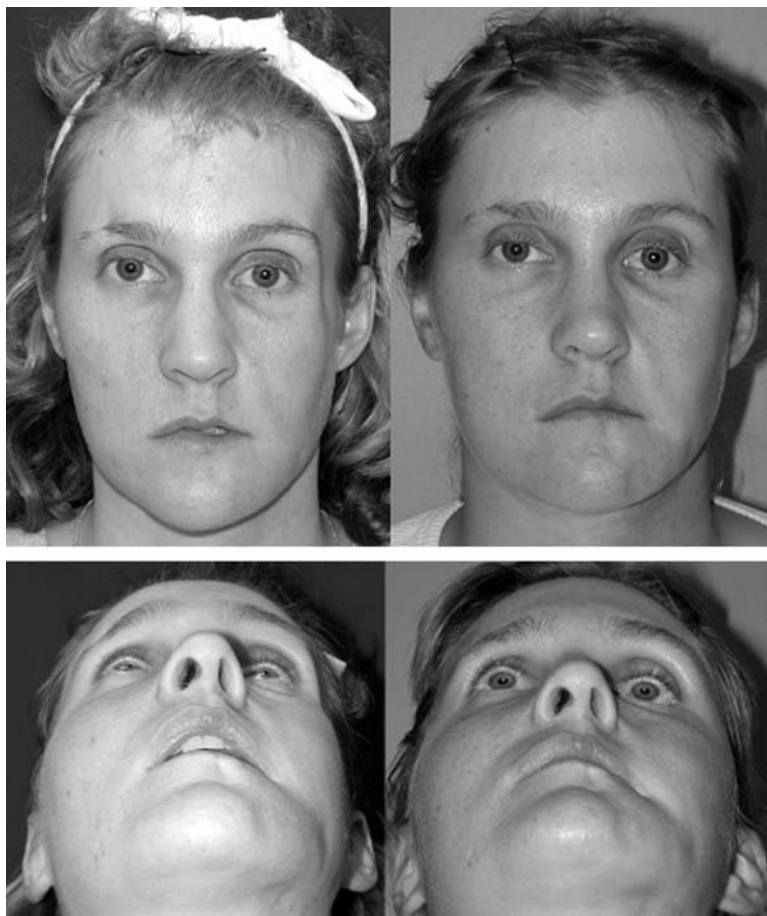
Temporal Hollowing

Post-traumatic temporal hollowing most commonly occurs following surgical elevation of the temporalis muscle with associated injury to the adjacent temporal fat pad, the muscle itself, and/or inadequate resuspension of the muscle to the temporal fossa at the completion of the procedure. This soft tissue contour deformity may be addressed with any of a number of currently available techniques including autologous fat grafting, insertion of acellular dermal matrix, and insertion of hydroxyapatite bone cement. The author currently prefers autologous fat grafting when the extent of the deformity and general patient circumstances allows it.

Although multiple stages of grafting are occasionally required, they can usually be performed in the office using only local anesthesia at both the donor and recipient sites.

Case 13

A 35-year-old patient with Romberg syndrome (shown for illustrative purposes) presented several years following parascapular free flap to left face performed elsewhere, with residual concave deformities of left temporal region and right supraorbital rim. Postoperative photos taken at 13 months following autologous fat grafting (12 cc to left temporal area, 1.5 cc to right brow region).



Temporal Hollowing: Case 13.

Maxillary and/or Mandibular Malunion with Malocclusion

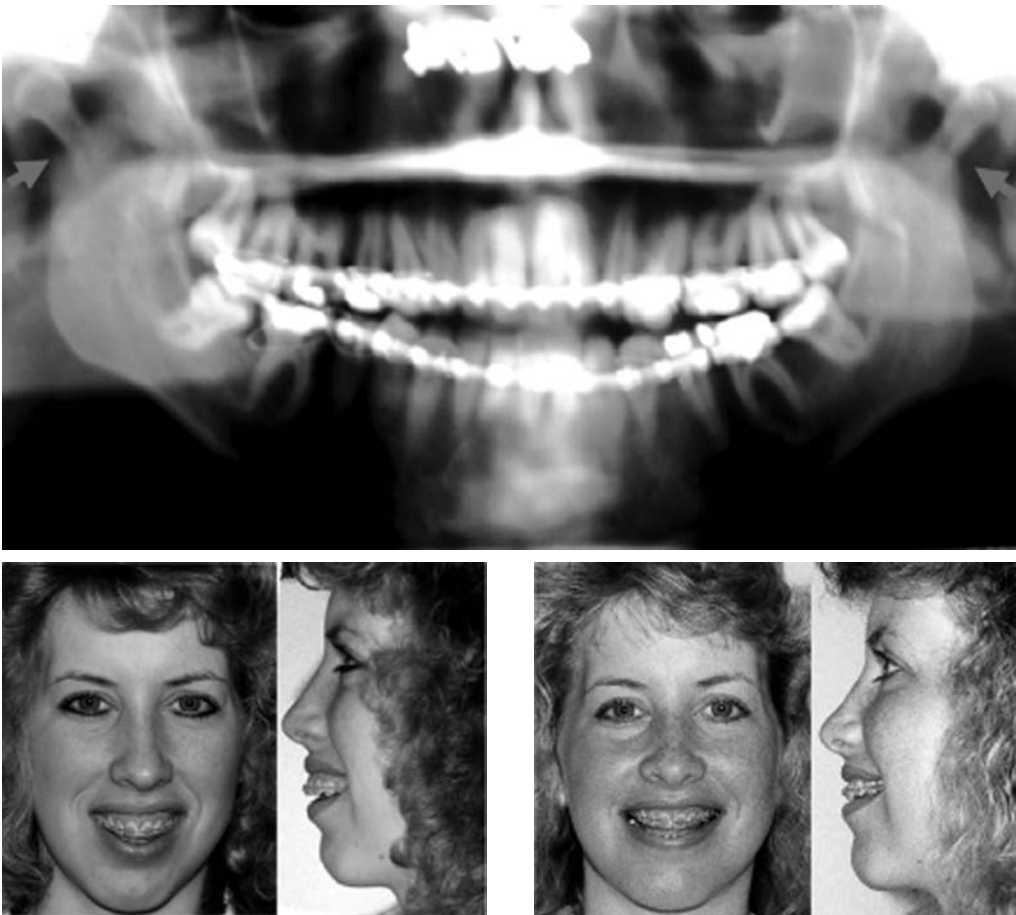
Malunited maxillary fractures most commonly are displaced posteriorly and cephalad. These fractures can be corrected with a Le Fort I osteotomy using fundamental principles of orthognathic surgery. Complex fractures may require segmental osteotomies. The acquisition of dental models is valuable, and allows one to perform model surgery and fabricate an occlusal splint preoperatively. Early referral and collaboration with an orthodontist is advisable. Examine the preoperative CT scan carefully to assess whether bone grafting will be necessary to support the midfacial vertical buttresses.

The correction of malunited mandibular fractures also can be best addressed using fundamental principles of orthognathic surgery. Fractures in the body and symphyseal regions can be corrected with refracture and repositioning techniques. Fractures in the

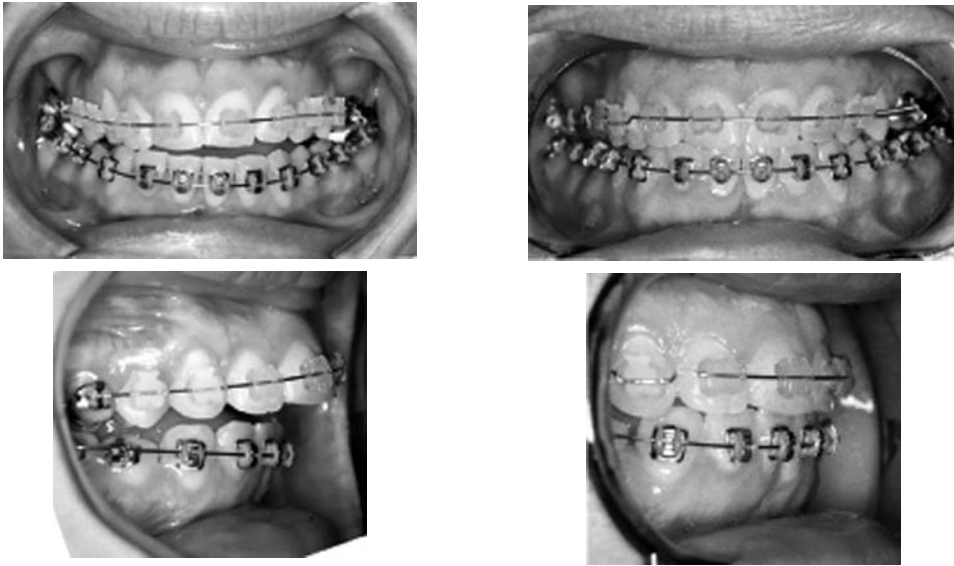
condylar region typically lead to ramal shortening and deviation of the mandible toward the side of the fracture with associated crossbite and chin point deviation. Ramal sectioning procedures are required to remedy the deformity; most commonly this will involve a bilateral sagittal split osteotomy. Bilateral condylar fractures will displace the mandible posteriorly and along with bilateral ramal foreshortening lead to an anterior open bite. With a prolonged assumption of this posture comes marked contraction of the masticatory muscles. These forces increase the risk of skeletal relapse greatly after ramal lengthening procedures in this scenario. A more appropriate reconstructive approach therefore is to perform a Le Fort I osteotomy with posterior impaction in order to allow for mandibular autorotation and correction of occlusion. An advancement genioplasty may be added to address the unfavorable aesthetic effect of the retruded mandible.

Case 14

A 24-year-old patient suffered bilateral subcondylar and symphyseal fractures resulting in condylar subluxation and bilateral ramal foreshortening. Initial treatment with intermaxillary fixation was inadequate and she presented several months later with an established post-traumatic anterior open bite and retrognathia. Preoperative assessment also revealed a prior underlying vertical maxillary excess. Definitive treatment at 27 months post-injury involved a segmentalized Le Fort I osteotomy with palatal division, allowing for both posterior widening and impaction of the maxilla. This was combined with bilateral sagittal split mandibular advancement osteotomies. Three-month postoperative photos are shown. *Source:* Courtesy of Dr. Rocco Addante, DMD, MD, Dartmouth-Hitchcock Medical Center.



Maxillary and/or Mandibular Malunion with Malocclusion: Case 14.



Maxillary and/or Mandibular Malunion with Malocclusion: Case 14.

Blast Injury with Composite Tissue Loss

The same general principles of craniofacial reconstruction apply to cases of massive composite tissue loss and destruction. One is called upon in these circumstances to employ the full range of technical modalities available to the plastic surgeon. These include, in particular, local and free tissue transfers and tissue expansion. The primary objective should be to address functional tissues such as soft tissue coverage of exposed skeleton, skeletal stability, protection of the globe, and oral continence. Only after the facial skeleton has been stabilized and restored to its anatomic volume should detailed reconstruction of normal soft tissue lining and coverage take place. Be aware that the reconstructive process may be confronted by ongoing and gradual resorption of the bony foundation of the face. In addition, the soft tissue envelope will undergo significant contraction followed by months of scar maturation after each individual intervention.

As the reconstruction proceeds, it is absolutely imperative that both the patient and the surgeon exercise a good deal of patience and creativity, along with a realistic expectation about the final outcome that may be achievable for these complex injuries.

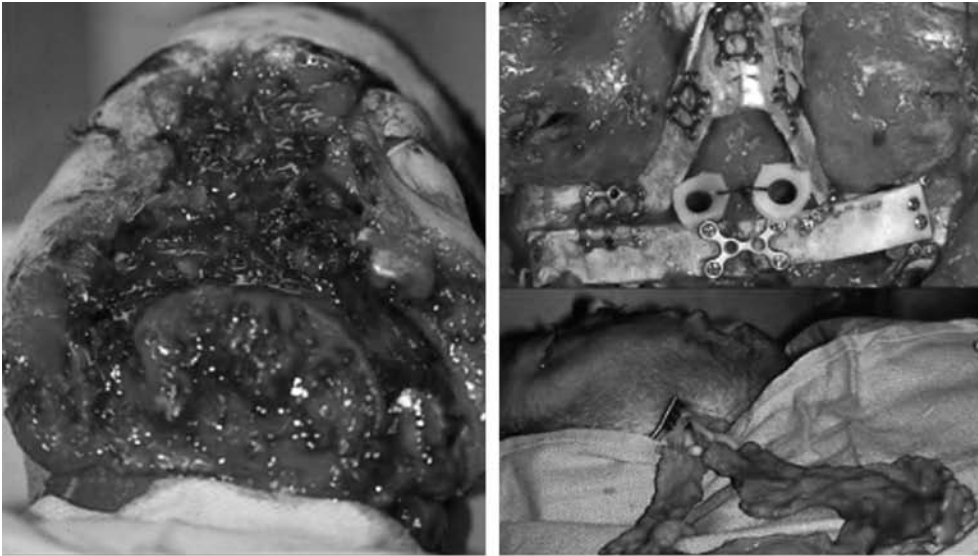
Case 15

A 19-year-old patient suffered a self-inflicted rifle blast injury. This resulted in massive soft tissue destruction along with bony loss and comminution of the NOE, zygomaticomaxillary, and central mandibular regions. The primary rate-limiting factor to this patient's facial restoration has been soft tissue lining as well as tissue perfusion and pliability. The author has performed numerous surgical interventions during the past seven years as reconstruction has proceeded in a stepwise fashion. The major procedures to date are listed:

- *Initial 3 weeks:* Irrigation, debridement, and wound closure followed by nasomaxillary skeletal reconstruction with split cranial grafts and omental free flap coverage
- *3 months:* Free fibular mandibular reconstruction (failed)
- *3½ months:* Radial forearm flap and mandibular plate reconstruction
- *10 months:* Split rib bone graft reconstruction of midfacial transverse maxillary arch
- *1 year:* Total forehead flap nasal reconstruction
- *21 months:* Left orbital cranial bone grafting (correction vertical dystopia and enophthalmos)
- *26 months:* Removal of mandible plate and free fibular mandibular reconstruction

- 36 months: Right temporal skin expansion begun
- 52 months: Cantilever bone graft to nose (rib), bilateral transnasal medial canthoplasties
- 54 months: Left temporal skin expansion begun
- 60 months: Forehead resurfacing and bilateral brow reposition to create upper lid sulci
- 73 months: Bilateral oral commissurotomy and correction of temporal hollowing with hydroxyapatite cement

Despite having achieved a stable midface and mandibular repair, substantial underlying skeletal resorption has occurred over time. The future plan of reconstruction includes further brow repositioning, bilateral medial canthal reposition, definitive nasal reconstruction, upper lip enhancement, and soft tissue revision of the chin.



Blast Injury with Composite Tissue Loss: Case 15.

Case 16

This 27-year-old patient sustained a self-inflicted shotgun blast resulting in significant injury to the right orbit (loss of vision in right eye), maxillary and mandibular comminution and bone loss, and major soft tissue defect to the mid and lower facial region including much of the mimetic musculature in that region.



Blast Injury with Composite Tissue Loss: Case 16.

- 2 days: Open reduction internal fixation of the mandible. This was covered with a pedicled pectoralis muscle flap. Skin grafts were applied to the muscle flap and to the large denuded midfacial region
- 1 month: Placement of bilateral 320 cc tissue expanders
- 7 months: Removal of tissue expander, debulking of muscle flap, removal of facial skin grafts, and resurfacing of upper and lower lip regions
- 30 months: Split cranial bone grafts to right orbit to correct enophthalmos

Future reconstruction will focus on nasal reconstruction, elaboration of the lips and oral vestibular sulci, and possible dental restoration. The greatest limitation is the lack of orbicularis oris sling and additional adjacent musculature which will restrict the ability to provide him with a mobile and competent oral stoma. He may be a candidate in the future for a functional free muscle reanimation procedure once the residual scar tissue has been excised or has matured, and improved oral lining has been provided.

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21 Craniomaxillofacial Prosthetics

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INTRODUCTION

Why a chapter on craniomaxillofacial prosthetics in a textbook dedicated to surgical procedures? Because, in spite of great advances in the surgical reconstruction of patients with craniomaxillofacial deformities, there are still many patients and particular facial defects which are better suited for prosthetic reconstruction. The ability to identify those patients and deformities early in the decision-making process will ultimately benefit both the patient and surgeon.

Achieving an optimal functional and cosmetic outcome for patients with congenital or acquired facial deformities is best addressed utilizing a skilled multidisciplinary team approach. In cases of head and neck cancer, a complete understanding of the disease process as well as the various treatment modalities, including radiation therapy, chemotherapy, surgical resection, and reconstructive options, is essential in order to determine the role for prosthetics. Extensive facial trauma involving a major loss or deformation of specialized tissue, such as the eye/orbit, the nose, or the ear, may necessitate the consideration of prosthetic reconstruction. In rare cases of severe congenital malformations, prosthetic reconstruction may be a consideration either primarily or after failed reconstructive attempts. An experienced team evaluation including the surgeon, oncologist, prosthodontist, restorative dentist, ocularist, and speech/language pathologist is beneficial in formulating an overall plan. Working together as a team will increase the chances of that patient returning to a more “normal” life.

CURRENT APPROACHES

Evolving surgical techniques in reconstructive plastic surgery have dramatically improved our abilities to restore both function and cosmesis to patients with defects of the zygoma, cheek, jaw, perioral region, bony mandible, alveolar ridge, esophagus, and the neck. This has been primarily a result of refinements in microvascular free tissue transfer. Attempts at reconstructing the specialized and more delicate structures of the eye and periorbita, the nose and the ear have been less than optimal and typically require multiple staged procedures and revision surgery. There are great examples of total ear and nasal reconstructions with excellent outcomes in selected patients. Great results can be achieved in children with congenital microtia and in older patients with cancer of the nose when essential adjacent tissues are healthy and available for reconstruction. There are also many examples of suboptimal results.

MAXILLOFACIAL PROSTHETICS—ADVANTAGES

In the right patient there are many advantages to prosthetic reconstruction, including:

- The ability to create an almost *exact* anatomical copy of the contralateral normal structure, i.e., eye, periorbital area, or the ear.
- The ability to better match both the color, size, and texture of the opposite normal side.
- The ability to change these characteristics as the patient grows, experiences skin color and texture changes, and ultimately begins to show signs of aging.

- The avoidance of multiple surgical procedures and associated risks, discomfort, down time, and complications.
- The ability to remove the prosthesis and check for recurrent local disease.
- Prosthetic reconstruction can be done in an outpatient setting with little or no anesthesia or pain.
- The ability, with rare exception, to deliver a superior cosmetic result in the aforementioned “specialized areas” compared to autogenous tissue reconstruction.

With recent attempts at “facial transplantation” and further refinements in this area it may someday be possible to reconstruct these specialized areas using carefully matched homologous tissue with better, more natural “living” tissue. The future in the area will be exciting.

PREOPERATIVE ASSESSMENT

Ideally, patients undergoing major ablative and/or reconstructive surgery would be presented first for a team evaluation. There are many factors to take into account when trying to determine whether autogenous or alloplastic reconstruction (or a combination of both) should be used. In patients with cancer, factors such as the age of the patient, the nature of the disease or deformity, anticipated course of treatment including the extent of surgical resection, type and duration of chemotherapy and/or radiation therapy, chance of recurrence and the psychosocial status of the patient all play a part in the overall assessment. The primary surgeon will most likely determine the extent of the resection and the resultant anatomic defect. Many of these patients are older, in poor health, and may not be able to undergo major reconstructive procedures even though they may provide the best reconstructive option. Maxillofacial prosthetics may be the best option for patients who are not considered good candidates for surgical reconstruction.

In patients whose deformity is a result of trauma, the decisions are less complicated because the extent of the defect is known after appropriate studies. The major decisions in this situation are more related to the age of the patient, the timing of reconstruction, and the specific areas to be addressed.

Congenital deformities are much more complicated. In addition to the specific deformity, factors to take into account are the age of the patient, timing of operations with regard to growth, sequencing of procedures so as not to have a negative effect of future growth or “burn any bridges” for later operations, and the psychosocial status of the patient. In general, autogenous reconstruction is preferred in growing children. Prosthetic reconstruction is sometimes a consideration when these techniques have failed or are not an option from an overall health standpoint.

If alloplastic or prosthetic reconstruction is considered to be an option, changes in the surgical procedure may be necessary to facilitate better fixation and contouring of the prosthetic device. The prosthetic specialist will be able to determine whether specific anatomical structures would be better to keep or remove to better accommodate the prosthesis. In many cases, preserving uninvolved parts may actually help or create difficulties in fitting the prosthesis. The principles of aesthetic subunit reconstruction also apply in maxillofacial prosthetics. While the idea of removing otherwise normal or unaffected tissue may be against most plastic surgeons nature, it may offer significant improvements in the overall cosmetic result.

If osteointegrated implants are to be utilized for fixation of the prosthesis, implants can sometimes be placed at the time of the initial operation, obviating the need for a separate operative procedure.

Patient motivation is an important factor in determining whether or not prosthetic reconstruction is the best choice. Optimally, candidates for prosthetic reconstruction should have access to other patients with similar defects who have chosen this technique. The opportunity to meet and talk with other patients can be an extremely powerful way to educate and motivate patients to better accept their disease process and allay fears about how they may fit back into social situations after prosthetic reconstruction. Most of all, this may give them more realistic expectations of what life may be like afterwards. They may, after seeing other

patients, decide to pursue surgical options for reconstruction. If actual patients are unavailable, good quality color photographs and actual prosthetic models should be available to show potential patients. These patients must be willing and able to remove their prosthesis for cleaning and be able to replace it. The placement of certain external prostheses may require considerable dexterity, making it difficult for some patients. If a patient is unwilling or unable to participate in this process, the prosthesis may find itself in a box on a shelf and a gauze used in its place.

Once a patient is found to be a good candidate for prosthetic reconstruction, a “full face” alginate impression is made. High resolution color negative prints or digital photographs are made for the documentation of critical anatomical landmarks and structures which may be absent after surgery.

MATERIALS

Materials used for facial prostheses typically include either acrylic, Silastic (room-temperature vulcanizing silicone), or a combination of both, depending on the area. This material must be similar in color, texture, softness, and flexibility to facial skin so as to look and feel as natural as possible (1).

METHODS OF RETENTION

Various types of retention options have been described including the use of medical adhesives, eyeglasses, magnets, osteointegration, and internal fixation retention (IFR).

Internal Fixation Retention

IFR is a relatively new and versatile way of attaching facial prostheses (2). This process involves making a hollow, seamless, and flexible “balloon” which can be compressed during placement but immediately returns to its previous shape when inside its cavity (Fig. 1A,B). The external prosthesis is attached to this and is secured in place by the perfect “fit” of the expanded balloon behind it. In cases of cheek, oral, or maxillary reconstruction, the balloon fits snugly into this cavity against nasal mucosa, thereby decreasing the loss of humidity at this interface. For this technique to work, the inner facial defect or cavity must be larger than the outer or external defect.

Osteointegration

Osteointegration can be used to provide rigid retention for orbital, nasal, maxillary, and auricular prostheses (Fig. 2A,B) (3). The main criteria for successful osteointegration is the presence of healthy bone in which to place the implants. A bar is typically attached to these abutments onto which the prosthesis is attached. This method may also be used in conjunction with IFR hollow balloon methods.

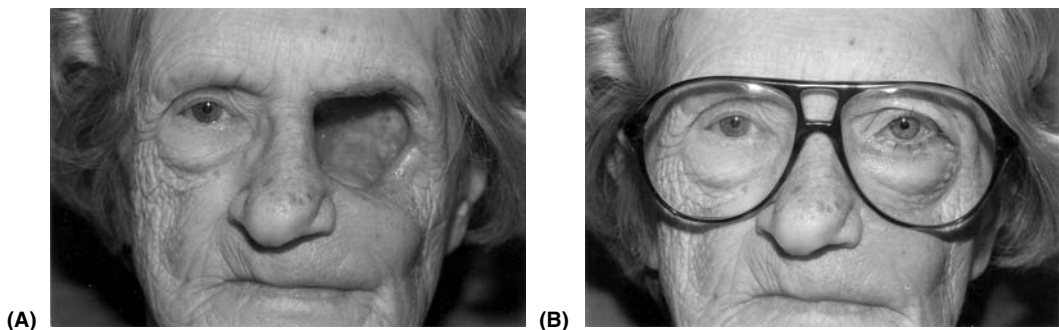


FIGURE 1 (A) Elderly patient with orbital defect after exenteration. (B) Same patient after internal fixation retention procedure.

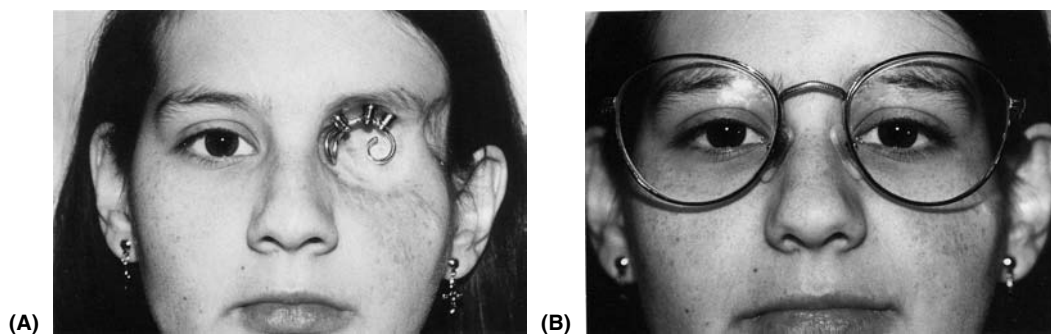


FIGURE 2 (A) Orbital/temple reconstruction utilizing osteointegrated implants and attached “bar” to which the prosthesis is securely attached. (B) Results showing excellent restoration of appearance.

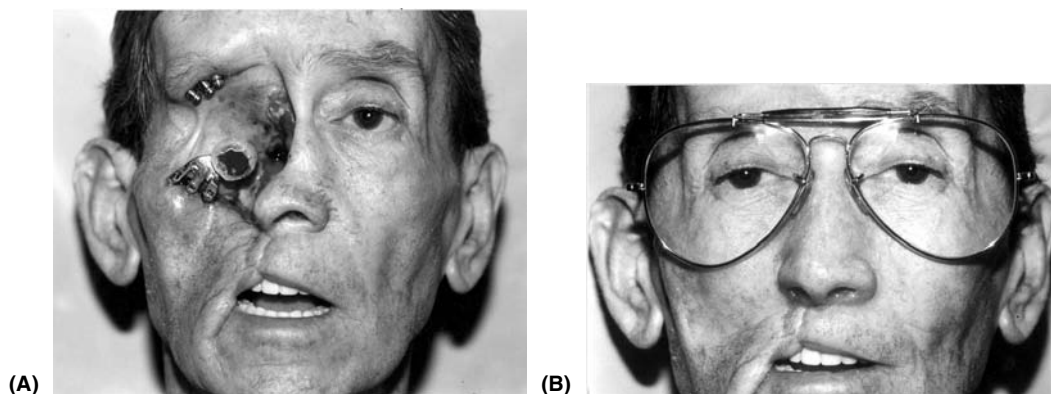


FIGURE 3 (A) Large complex deformity of the orbit, temple, nose, and cheek/maxilla. (B) Results utilizing a combination of magnets and osteointegration for retention.

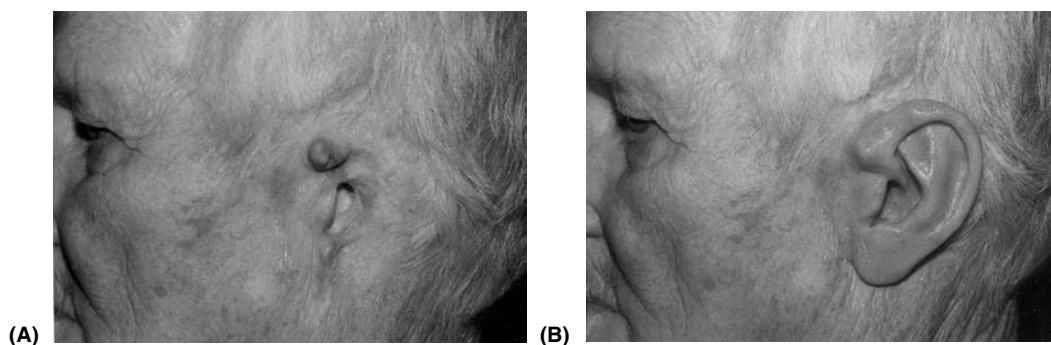


FIGURE 4 (A) Ear deformity secondary to cancer resection (note the smooth surface behind the canal for ear attachment). (B) Prosthetic ear with medical adhesive for retention.

Magnetic Retention

Extremely small, yet strong magnets, such as a neodymium magnet, can be used by themselves to attach to other magnets or ferromagnetic materials on the prosthesis to retain its position in the face. They can also be used in combination with osteointegration and IFR for maximum versatility and retention (Fig. 3A,B).

Medical Adhesives

Medical adhesives were used frequently in the early days of maxillofacial prosthetics. Allergic reactions occurred not infrequently, resulting in severely irritated skin, and making it difficult to wear the prosthesis. Many of the early auricular prostheses were held in place with adhesives (Fig. 4). In active patients, the prostheses could come off, especially with sweating or swimming, leading to embarrassment and noncompliance. With more advanced fixation techniques, the use of medical adhesives has become less common.

SPECIFIC AREAS

Matrix Prosthesis

With extensive resections of tumors of the face which extend back into the facial sinuses, nasal cavity, nasopharynx, palate, or maxilla a very large defect can result. In these cases a single balloon IFR may not be adequate. A matrix prostheses consisting of multiple interlocking balloons may simplify retention of the prosthesis (Fig. 5A,B,C). These are placed in the defect one piece at a time until it is totally filled.

Eyes

If a patient undergoes a simple enucleation, this is filled at the time with an orbital implant and covered later with a scleral shell, which can be matched to the other eye. If the extraocular muscles are intact, movement of the eye is possible resulting in a very natural result.

Orbit

With an exenteration, an orbital prosthesis is needed for optimal results. Good lining of the cavity is needed so any structures that can be safely left should be left (i.e., eyelids, eyelid skin, etc.). The orbital prosthesis is made and attached using IFR and/or osteointegration techniques. Regardless of the type of fixation used, a hollow eye socket is created. A custom, impression fitted hand painted artificial eye is then placed into this socket in central gaze. Properly fitted and correctly positioned eyeglasses are used to protect the only sighted eye and to camouflage the edges of the prosthesis (Fig. 6).

Nose

Excellent results in nasal reconstruction have been achieved by numerous authors utilizing local and adjacent tissues (4). In cases where these tissues are not available or the patient is not otherwise a good candidate, prosthetic reconstruction can achieve excellent outcomes. Communication with the primary surgeon is critical here in determining which tissues to resect or leave. If the majority of the nose is to be removed and prosthetic reconstruction is anticipated, it is easier and aesthetically beneficial to perform a total rhinectomy. Leaving parts of the ala or heminose may actually interfere with fixation and be difficult to work around. From the standpoint of aesthetic subunits, it is better to replace the entire nose (Fig. 7A,B). In the coauthor's experience, IFR (as discussed previously) is the fixation method of choice for nasal alloplastic reconstruction.

Auricular Prosthesis

In cases of advanced skin cancer of the ear where greater than half of the auricle has to be removed, prosthetic reconstruction can provide excellent results. Patients with congenital

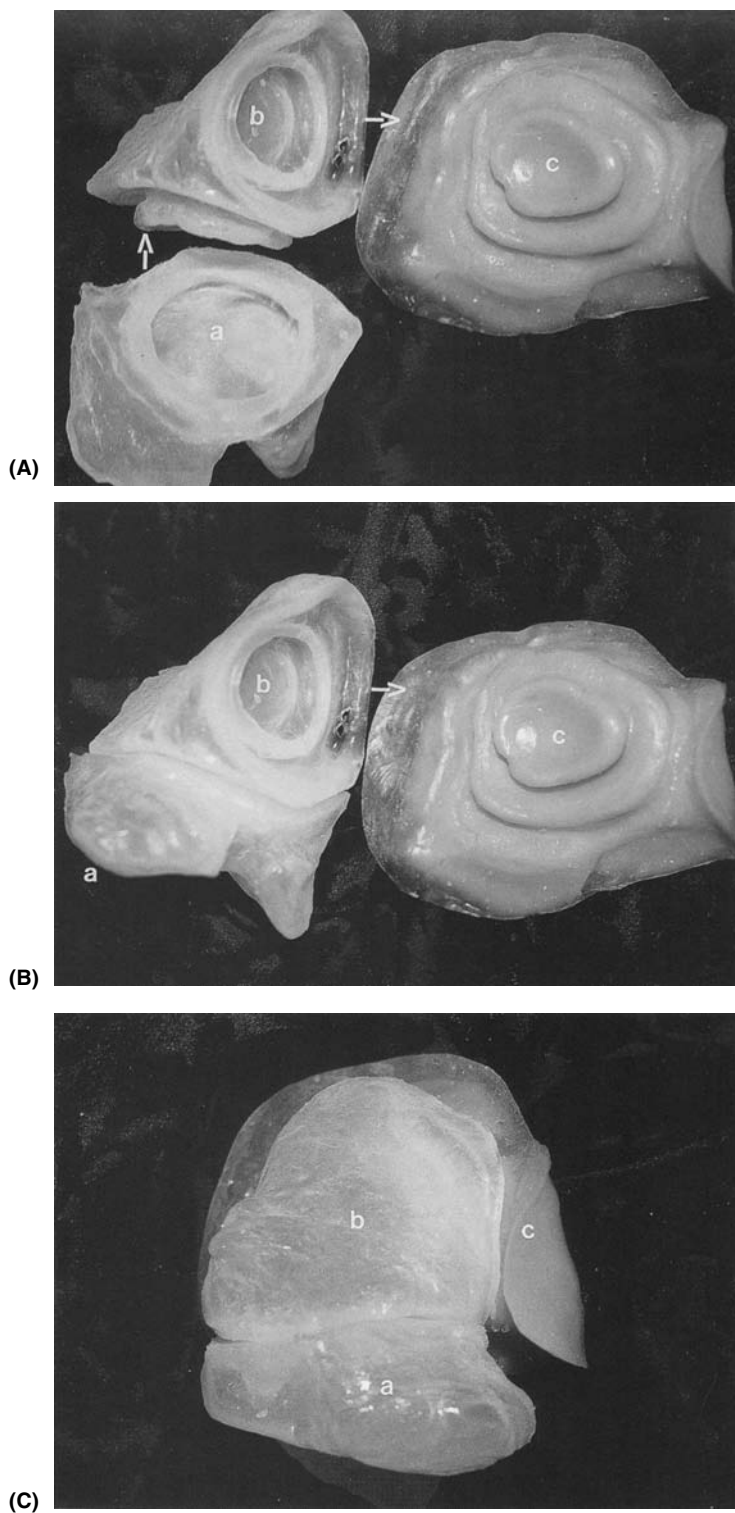


FIGURE 5 (A) Matrix prosthesis for very large, complex defects (the smaller pieces a, b, and c, sequentially interlock to build and fill the defect). (B) Two pieces joined to the third. (C) Complex matrix prosthesis.

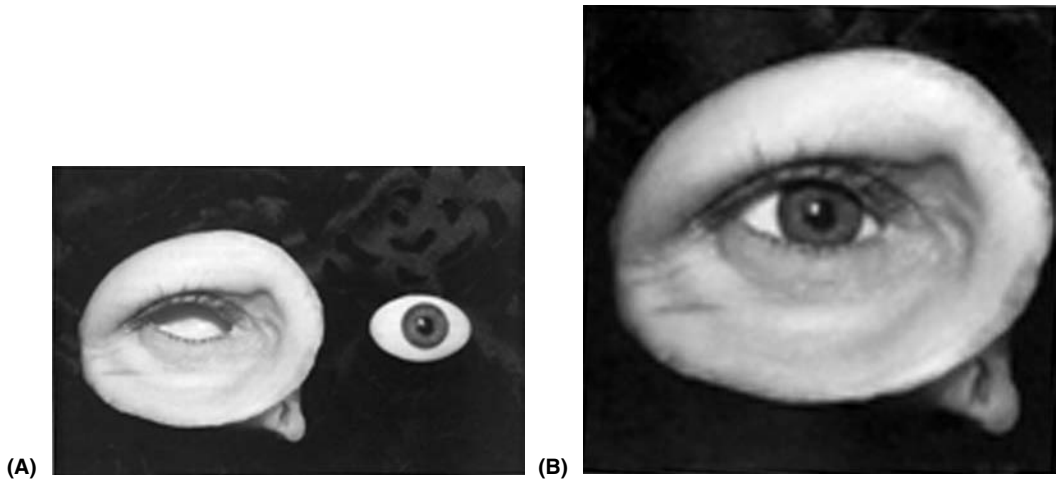


FIGURE 6 (A) Orbital prosthesis with empty socket and separate ocular prosthesis. (B) Completed two piece reconstruction.

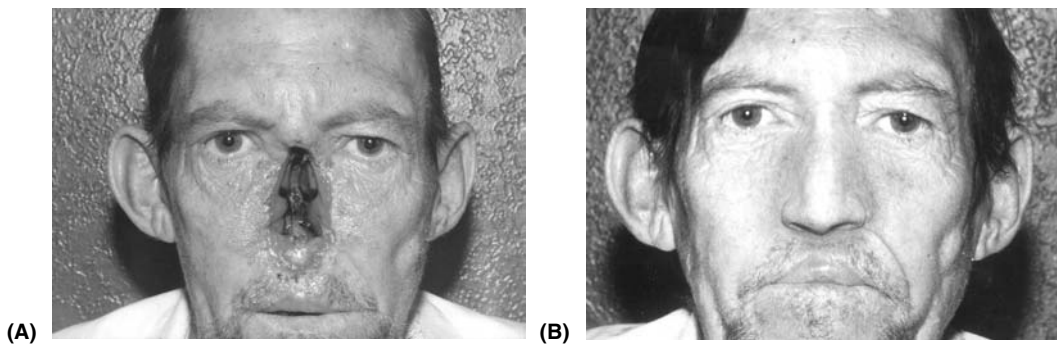


FIGURE 7 (A) Patient after total rhinectomy. (B) Completed alloplastic reconstruction retained by internal fixation retention.



FIGURE 8 (A) Complex deformity of the temporal area and absent auricle (note three osteointegrated implants and retention bar). (B) Completed, rigidly fixed prosthetic ear reconstruction.



FIGURE 9 (A) Patient with massive trauma to the face. (B) Resultant defect with multiple anatomic areas involved. (C) Fixation in place (implants and internal fixation retention). (D) Final result.

auricular deformities such as anotia or severe hemifacial microsomia may also benefit from prosthetic auricular reconstruction when autogenous tissue reconstruction has failed (5). Communication with the primary surgeon is critical in these cases prior to resection. Leaving “tags” of ear such as the lobule, parts of the helical rim, or concha are not beneficial. Total auricular reconstruction provides a better cosmetic outcome than a partial one. The tragus is the only anatomical part that may be of benefit aesthetically. Although external ear prostheses may be attached with medical adhesive, the optimal method is with osteointegration. This can provide much greater stability, especially with active young children or adults (Fig. 8A,B).

Combined Nasal–Orbital–Facial Prosthesis

Some cases involve massive tissue loss to the face and underlying structures, such as after a gunshot wound or a large tumor resection. These can leave not only external facial deformities but large cavities where they open into the sinuses. In many instances it is desirable to fill this space. Muscle or myocutaneous flaps, whether free or pedicled, can be used to accomplish these goals. In some patients, there may be a need to visualize these cavities later to look for recurrence. In this case, a combined facial prosthesis may be the best option (Fig. 9A,B,C,D). Fixation using IFR allows the cavity to be sealed while the prosthesis is in place and easily examined when the fixation balloon has been removed (Fig. 10) (6). In the case of a maxillary defect, the IFR balloon can serve as an obturator to decrease the associated nasopharyngeal

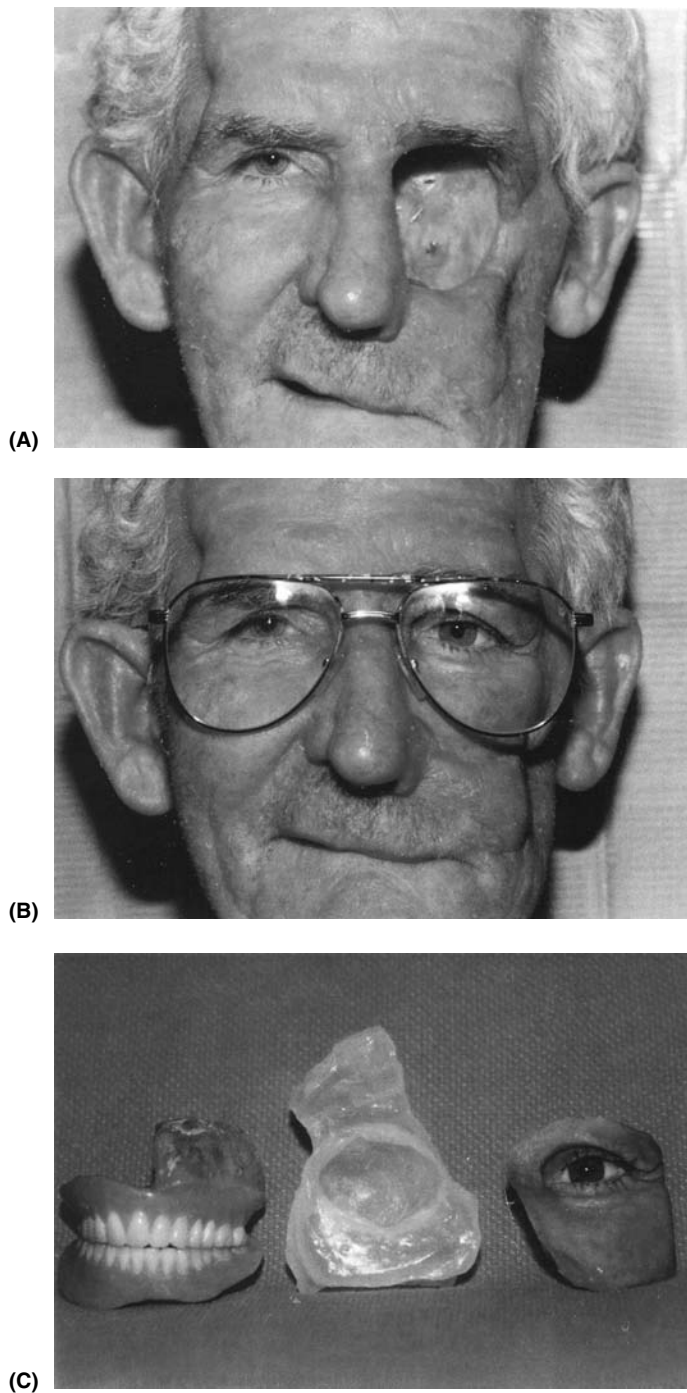


FIGURE 10 (A) Complex oculo/orbital/maxillary defect. (B) Result. (C) Component prostheses, removable for cleaning and examination of the area for recurrence.

insufficiency and hypernasal speech. As with all of these cases, good communication with the surgeon preoperatively and sometimes even intraoperatively can maximize results.

SUMMARY

Severe facial deformities often result from head and neck cancer ablation, trauma, and severe congenital malformations. Many of these are best addressed using surgical techniques involving the movement of local, regional, or distant tissue. Advances in free tissue transfer have had a major impact on reconstruction, especially mandibular reconstruction. When more specialized areas of the face such as the eye, periorbital, nose, ear, or combinations of these are involved, prosthetic reconstruction should be considered. Excellent functional and cosmetic outcomes can be achieved utilizing a combined team approach to the patient. Advances in fixation techniques, especially utilizing osteointegrated implants and/or IFR, have made prosthetic reconstruction even more effective. Surgeons who participate in the care of these patients should be aware of the benefits and uses of maxillofacial prosthetics and the role in can play in maximizing the overall restoration process.

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Evaluation, Management, and Avoidance of Complications in Craniofacial Surgery

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INTRODUCTION

Along with the enormous challenges and rewards encountered in the field of craniofacial surgery, there is also the potential for devastating complications. Several authors report an overall complication rate among craniofacial procedures between 14% and 22%, with a mortality rate around 1% (1,2,3). The more common complications described include bleeding, infection, and continued postoperative deformity requiring further intervention. Before addressing complications specific to certain procedures, several basic risks are applicable to all craniofacial procedures.

Airway

Elective craniofacial procedures introduce a risk of inadvertent extubation resulting from frequent turning of the head and neck during surgery. For all cases, proper care for the endotracheal tube should be of paramount importance. With oral intubation, the tube should be well-taped in the midline or off to one side depending on the procedure. With nasal intubation, a suture should be placed around the tube on a short "mesentery" and sutured once to the caudal nasal septum and once to the forehead behind the hairline. Gauze sponges beneath the tube will prevent injury to the skin and subcutaneous tissues. The tube should be positioned to minimize traction and kinking. Superior traction on the tube should be avoided to prevent injury to the alar rim during a long procedure. Aspiration of blood and saliva should also be avoided. Many smaller diameter tubes do not have a cuff to prevent aspiration. In all cases, a moistened throat pack should be placed in the posterior pharynx to prevent aspiration of blood and saliva.

Postoperatively, the decision to leave a patient intubated overnight should not be taken casually. In small children with craniofacial syndromes and aberrant anatomy who require shorter, uncuffed endotracheal tubes, subtle changes in the position of the head and neck may dislodge the tube into the esophagus or proximal oropharynx. Examination of the chest should always be performed and a confirmatory chest x-ray should be taken whenever a patient is moved from one location to another. Clinical examination alone is not always reliable and appropriate monitoring should also be utilized.

Concomitant injuries are frequently encountered in patients with craniofacial trauma. Significant injuries to the brain, spinal column, chest, abdomen, or extremities trauma take precedence and may be fatal if unrecognized. Evaluation of a patient's airway, breathing, and circulation should always precede any intervention to repair fractures of the craniofacial skeleton. A thorough primary and secondary survey should attempt to identify these urgent injuries. Injury to the cervical spine should be ruled out with appropriate radiographs prior to operative intervention. These may include plain films, flexion-extension images, and/or CT scans.

Airway obstruction is a potential lethal complication of both injury to and intervention in the head and neck. Aside from generalized bleeding, severe nasal fractures may completely occlude the upper air passages and make respiration difficult. Bilateral mandibular fractures

with posterior displacement of the anterior segment and base of an edematous tongue may lead to worsening airway compromise and ultimate asphyxiation.

With any question regarding the adequacy of ventilation, the airway should be secured either with an endotracheal tube, if possible, or a tracheostomy. Caution should be exercised in placing an endotracheal or nasogastric tube because blood may turn a routine intubation into a difficult ordeal. Also, comminuted fractures of the posterior facial skeleton may produce pathways into the calvarial vault.

Hemorrhage

Bleeding in the head and neck may be challenging to identify and control due to anatomic limitations. The tissues are generally well-vascularized and bleed promptly with intervention. In addition, the surgeon is often working in areas not directly visualized and not easily exposed, as is the case with the posterior nasopharynx and the pterygomaxillary fossa. In infants and young children, careful recognition of bleeding is important since a small loss may represent a large fraction of the total circulating blood volume. In general, approximately 8% to 9% of an infant's weight is equal to his circulating blood volume. Replacement should be made available if blood loss is anticipated.

Maneuvers to minimize bleeding are important and should not be overlooked. Infiltration with local anesthetic agents should be routine and ample time should be allowed to exert their effect. As with profuse bleeding during partial removal of a tumor that slows considerably once the tumor is fully excised, bleeding may be vigorous during performance of osteotomies but will often abate with final down-fracture. If significant bleeding is encountered, compression of the disparate segments and packing the area with epinephrine-soaked pledgets may be helpful.

Infection

Infection is seen in roughly 5% of craniofacial procedures. It is most frequently encountered in the acute postoperative period after prophylactic antibiotics have been discontinued. The oral cavity, sinuses, and periodontal structures provide an adequate source of contaminating organisms. The most commonly offending organism is *Staphylococcus*. Other notable offending agents include *Streptococcus* and *Pseudomonas*. Interestingly, *Pseudomonas* was noted to be the most common offending organism in a series of patients treated in Australia over a 10-year period (4). Contributing factors included the presence of a tracheostomy and longer operating times. Residual dead space following frontal advancement was reasoned to be a sanctuary to infecting organisms from the respiratory tract transferred from a tracheostomy site by ventilatory forces. Of note, the adults in this series had a higher rate of infection than did the children.

The risk of infection is compounded by injury to the surrounding blood supply, which impairs blood flow to a fracture site. Acute infection is more commonly an infection of the soft tissues. Experience with fracture management has improved infection rates with elective procedures. In repairing fractures adjacent to sinus cavities, there is the potential for contamination. With regards to the frontal sinus, removal of the posterior sinus wall and obliteration of the frontonasal ducts is required with involvement of the posterior table of the sinus.

Infection following repair of midfacial fractures may occur in patients with preexisting sinus infection or concomitant poor oral hygiene. Presenting complaints typically appear within the first one or two weeks postoperatively as tenderness, erythema, swelling, and/or discharge of malodorous fluid into the oral cavity. Prevention involves the liberal use of parenteral antibiotics and perioperative oral hygiene. Oral rinses, such as Peridex, should be used pre- and postoperatively. Once in the operating room, a thorough cleaning of the oral cavity should be performed. The teeth should be brushed using a dilute solution of iodine-povidone and peroxide. Further injury to the tissues should be minimized by opening the mucosa with an adequate cuff on either side for reclosure at the conclusion of the procedure.

Submucosal muscle should be left with its pedicle so as not to create large devascularized segments. On the maxilla, this involves dissection beneath the mucosa as inferiorly as possible to protect the lip and cheek elevators. On the mandible, this involves dissection as superior as possible to protect the mentalis muscle. Only as much periosteum should be stripped off the bone as is needed for exposure, reduction, and repair. Often, the soft tissue attachments of free-floating segments of bone may be preserved during exposure and fixation to enhance the vascularity of the segment.

Fractures of the mandible more commonly result in infection than do those of the maxilla. The normal flow of lymph with the mandible drains inferiorly to the inferior dental canal and then out the mental foramen and into the submandibular lymph nodes. Fractures of the mandible disrupt the normal flow of lymph and allow organisms from within the canal to inoculate the fracture site preferentially (5).

Prophylactic antibiotics are recommended when incisions are made in the oral cavity, intracranial procedures are planned, or bone or cartilage grafts are employed. They should be started before the induction of anesthesia, approximately 20 minutes prior to the incision, and not once the case is underway. Many centers use a first or second-generation cephalosporin for broad-spectrum antibiotic coverage. For penicillin allergic patients, clindamycin, which covers many gram positive and anaerobic organisms, is a valuable alternative.

Once infection is established, the diagnosis can usually be made by history and physical examination alone. Treatment follows basic surgical principles and involves opening the wound, adequate removal of all nonviable and foreign material, copious irrigation, and secondary closure with a drain. Chronic infection more commonly follows a comminuted fracture with avascular necrosis of the bony fragments. Extensive osteomyelitis requires debridement of bone back to healthy, vascularized tissue. Gaps in the bone will later need to be filled with autogenous bone graft after the infection has cleared. Temporary stabilization may require placement of external fixation.

Malunion and Nonunion

Malunion usually results from failure to either adequately reduce disparate fracture fragments or not establishing adequate bone-to-bone contact. With bilateral fractures, posterior displacement of the segments often needs to be corrected by restoring the correct anterior projection of the face. This begins by identifying the points of fracture and reducing each. Accurate reduction and fixation of the zygoma will restore the correct sagittal position of the malar complex. Vertical height of the maxilla is restored by accurately reducing the medial and lateral buttresses.

Nonunion of maxillary bone is rare. It often presents with continued pain and mobility across the osteotomy site. Causes include improper initial alignment, postoperative movement of the segments, and infection at the site of the injury. Adequate healing requires the ends of the bone to be properly aligned, free of nonviable tissue, and an appropriate period of immobilization.

In the case of a mandible fracture that heals with a nonunion following treatment with intermaxillary fixation, the means of fixation (such as arch bars or suspension wires) needs to be evaluated. Improper fixation of the teeth to the arch bars will result in movement between the jaws and poor fixation. After tightening the arch bars, it is useful to grasp each bar with a clamp and gently rotate the head (with all cervical spine precautions taken as necessary) to determine if there is any movement between the teeth and the hardware. The entire jaw should move with the bar. If loose, the wires need further tightening or reinforcement with extra points of fixation. It must be remembered that wires only loosen with time.

Treatment of nonunion requires debridement of the fibrous ends and placement of an autogenous bone graft or vascularized bone flap if large. This may be done through an external approach to minimize the risk of contamination. Discrepancies of occlusion may require fabrication of dental models and an intraoperative occlusal splint to restore the proper intermaxillary relationships.

CLEFT LIP REPAIR

Dehiscence

Prevention of dehiscence following cleft lip repair begins with choosing the appropriate initial management for the patient. For children with an incomplete cleft solely of the lip or a complete cleft with well-aligned palatal arches, the tension across a repair may be minimal and the risk of dehiscence will therefore be small. Repair may be undertaken at three months of age with minor concern for postoperative separation. However, infants with a widely spaced complete cleft of the lip and palate will likely require preoperative intervention. This serves two major roles. One, the improved positioning of the palatal arches will lessen the tension across the repair and minimize the risk of dehiscence. Second, the diminished tension across the repair, combined with the ability to mold the nasal cartilages, should improve the aesthetic outcome of the repair (6,7). The development and continued improvement in presurgical nasosilveolar molding has improved the palatal alignment and decreased tension across the lip in many cases of unilateral and bilateral clefts of the lip and palate (8,9). In the past, lip adhesion was often used to impart tension across the soft tissue envelope of the face in hopes of moving the disparate palatal arches closer together.

Intraoperatively, tension across the lip may be minimized by adequate release of the soft tissue elements tethered on the maxilla. Postoperatively, a well-adherent tape dressing should be used in conjunction with arm restraints. Feeding protocols vary from center to center. Once dehiscence has occurred, it is best to allow any inflammation or cellulitis to subside before attempting a secondary repair.

Poor Result

The poor result in cleft lip surgery is easily recognizable by even the untrained eye. It may be manifest as a noticeable suture line, a short lip, a jagged mucocutaneous junction, and/or a deficient tubercle. Accurate caliper measurements and tattoo markings should be done prior to injection of anesthesia and before any incisions are made. The injection of local anesthesia blanches key landmarks and incisions, once made, are not retractable. The incisions should bisect any tattoo points so that a portion remains visible for use during the procedure to guide the repair.

Postoperatively, a vertical lip scar will tend to contract, raising the high point of Cupid's Bow asymmetrically. This process may be ameliorated by a small Z-plasty just above the vermillion edge as described by Onizuka (10), among others. It serves not only to break up the linear nature of the scar but also to provide length to the philtral column on the cleft side.

The choice of suture material may play some role in the final outcome but varies widely from surgeon to surgeon. Sutures should be of the smallest reasonable caliber, be placed without tension, and removed within a relatively rapid period of time so as to minimize scarring. Most cutaneous sutures used today are either 6-0 or 7-0 in caliber and either nylon, which must be removed, or plain gut, which dissolves with time. Sutures placed along the reconstituted philtral column should carefully evert the skin edges to mimic the normal contralateral side. A flat suture line in this area is unnatural.

Revision of a jagged border at the mucocutaneous junction may be performed at a later date, often with a small Z-plasty to realign the edges and lengthening the short lip. Minimal discrepancy between the cleft and non-cleft sides may be treated with an elliptical excision of the scar and linear closure of the defect. More significant shortening requires recreation of the component parts and further rotation and advancement of the medial and lateral lip elements.

Deficiency of the tubercle may be addressed secondarily by the lateral advancement of neighboring mucous membrane as described by Kapetansky(11). An ellipse of tissue is designed in the lateral lip element and based superiorly within the substance of the lip. It is inserted into the adjacent area of deficiency and the donor site is closed primarily in a V-to-Y fashion.

In the bilateral cleft lip, symmetry is key and both sides should be repaired at the initial setting. Closure of one side will impart unequal tension on the prolabium and premaxilla and worsen the defect of the unrepaired side. An unsightly result has resulted from the desire to

import tissue into the central portion of the lip to make up for the inherent deficiency. The surgeon must accept whatever prolabium is present and not introduce portions of the lateral lip elements as a way of increasing the height of the philtrum. The mucosa of the central prolabium should be turned over for lining and sutured to the corresponding tissues of the lateral lip elements.

CLEFT PALATE REPAIR

Errors of Speech and Incomplete Facial Growth

Repair of the palate focuses on function as well as form. Much has been written about the timing of palatal closure (12,13). A well-crafted repair may have significant adverse sequela if performed either too early or too late. Some authors have advocated delayed palatal closure to avoid interfering with facial growth (14). Others have disputed this approach (15). Eight to twelve months of age is considered a relatively safe time for repair but no single criterion should be relied on to determine the proper timing of palate closure.

Postoperatively, the short and/or scarred palate will demonstrate little movement and often contribute to velopharyngeal incompetence. Identification of the short palate prior to formal cleft palate repair may warrant performance of a double-opposing Z-plasty (16,17) for closure rather than a straight-line closure with an intravelar veloplasty. Later maxillary hypoplasia in the setting of repaired cleft of the lip and palate may be addressed at the time of skeletal maturity with formal orthognathic surgery.

Fistula and Dehiscence

Avoidance of fistula formation begins with adequate closure under minimal tension. Postoperative instructions should be clear to avoid placing straws, spoons, or suction catheters into the oral cavity. Suctioning in the operating room or recovery room should be done either by the operating surgeon or by caregivers who are intimately aware of the operative procedure and its potential complications. Arm restraints are suggested postoperatively for two weeks, especially in patients with greater motor development and more anterior clefts. Again, feeding protocols will vary from center to center.

Once a fistula is recognized, it is best to avoid returning to the operating room in the immediate postoperative period. It may be advisable to start the patient on a course of oral antibiotics and rinses to limit any contributing infection or inflammation. Observation for a period of six months or longer avoids operating on inflamed mucosal tissue. It also allows smaller fistulae to close spontaneously. Those located at the junction of the hard and soft palate contribute less to hypernasal resonance than do more anterior fistulae. Obturation of a fistula with chewing gum may be used to judge its contribution to errors of speech.

Again, delayed closure must be achieved without tension. The healed mucosa covering the palate has little elasticity. Local flaps must be widely undermined so that they may be transposed with minimal tension across the repair. Delayed repair is best performed as a two-layered closure with a smaller medially based flap from one side of the defect turned over for nasal lining and a larger flap from the contralateral side advanced medially for oral lining.

CLEFT ALVEOLAR REPAIR

Mucosal Dehiscence

Repeated trauma to the oral mucosa can result in problems with wound healing. Mucosa lacks the strength and elastic properties of skin and therefore must be handled with care. When incising the mucosa, a scalpel should be used to create a viable edge and care should be taken around the teeth to protect the interdental papillae. Logic dictates that closure should be under minimal tension and be water-tight to avoid contamination of the underlying bone fragments. A periodontal dressing (Coe-Pak) may be applied to further protect the repair in the immediate postoperative period.

Infection of the Donor Site

The combination of a sterile donor bone graft site and a colonized recipient site puts the former at risk for infection following harvest. This may be minimized by copious irrigation and careful separation of the various instruments. Forceps and the like that are used in the oral cavity should not then be used in either the harvest or closure of the hip wound. In addition, the technique by which bone graft is harvested may also minimize the risk of infection. Recovery of cancellous bone graft through a small stab incision over the anterior superior iliac crest using a percutaneous bone biopsy kit exposes less of the iliac crest, which is thought to improve postoperative ambulation and decrease the risk of infection.

Bone Resorption

Resorption of bone is affected by the adequacy with which the maxilla is sealed off from the nasal vestibule and oral cavity. The graft is usually not fixed to the margins of the cleft and may be lost if mucosal closure is inadequate. Similarly, the soft tissues along the inner margins of the cleft should be preserved to reconstruct the nasal floor with adequate substance to create seal through which no bone graft will be compromised. The oral closure must be equally adequate to prevent contamination of the graft by saliva and oral fluids.

INTRACRANIAL SURGERY

In treating children with anomalies of the craniofacial region, the surgeon benefits from a multidisciplinary approach, which traditionally involves any or all of the following: a plastic surgeon, a pediatric otolaryngologist, an oral surgeon, a pediatric dentist and orthodontist, and a speech/language pathologist. Other specialists, including an audiologist and psychologist, may also evaluate each child. Examination of the child with plagiocephaly should identify ridging, suture patency, absence of bony continuity, areas of fullness or hollowing, visual acuity, and age-appropriate neurologic evaluation. Radiologic evaluation may include plain films of the skull and/or CT scanning which permits reconstruction of the images in three dimensions.

Injury to the Brain, Meninges, and Eyes

Surgery to remove the calvarial vault and remodel the bones around the orbits puts vital structures, such as the brain, meninges, and eyes at risk during burr and osteotomy creation. During exposure, the dura may be tightly adherent to the undersurface of the skull. Initial burr holes should be only deep enough to contact the dura. Further enlargement of the openings may be done with a curette or rongeur to minimize injury from the burr. Extradural tunnels should then be carefully developed to provide unimpeded access for the calvarial drill (Fig. 1). Dissection across the sagittal sinus should occur last to allow rapid entrance into the cranial vault should bleeding occur. Once all the osteotomy sites have been created, elevation of the bone segments should take into account areas over the fused sutures where invagination of the sinus (Fig. 2) may produce a tight attachment and lead to inadvertent laceration.

Once the cranial vault is entered, all osteotomies should be under direct vision. Soft tissue opposite the point of the drill or saw should be protected with a malleable retractor (Fig. 3). This is especially important around the globes, which lack a dural covering. Continuous irrigation should be used to minimize thermal injury to the edges of the bone flaps.

Meningitis

Meningitis may occur as a consequence of exposure of the intracranial contents from the nasal cavities and ethmoid sinuses. Careful irrigation, drainage, and perioperative antibiotics may minimize the risk. Kawamoto (18) reported excellent results with midfacial advancement procedures in patients with Apert's and Crouzon's, but noted a higher complication rate in those patients with ventriculoperitoneal shunts, possibly due to inability of the soft tissues to



FIGURE 1 Dissection of the dura off the underside of the cranial vault through burr holes.

accommodate the increased volume of the cranial vault and fill the extradural space following the advancement. In advancing the midface, consideration should be given to whether or not concomitant advancement of the cranial vault is required. If not, a subcranial Le Fort III may be performed that does not violate the calvarium.



FIGURE 2 Synostotic region of the sagittal suture demonstrating growth of bone around the sagittal sinus.



FIGURE 3 Protection of the underlying soft tissues from inadvertent drill or osteotome injury with a wide, malleable retractor.

Globe Injury

Injury to the globe may be prevented by meticulous dissection within the orbit and careful preservation of the periorbital soft tissue envelope around the globe. In rare instances, such as with a fronto-orbital advancement, it is necessary to open the periorbita. This should be done with a scalpel and gentle spreading of a clamp or scissors. Injury to the more posterior optic nerve is a more severe injury that may be prevented by maintaining a safe distance from the orbital apex, which lies approximately 45 mm from the rim along the floor. As a general rule, safe dissection should proceed no further than is necessary and rarely deeper into the orbit than 35 mm. The posterior shelf of a typical orbital floor fracture is no further than the posterior wall of the maxillary sinus. Introduction of a blunt instrument into the sinus via the floor should locate the posterior wall and identify a safe distance for dissection and placement of bone graft for repair.

Undercorrection

Undercorrection may result from incomplete initial craniectomy and/or reconstruction. With extensive, multi-sutural involvement, complete correction of the deformity may not be possible at the initial procedure. Additionally, the presence of a ventricular shunt may impair adequate single-stage correction. For sagittal synostosis, simple strip craniectomy, as initially described by Land (19) and Lannelongue (20) in the 1890s may not be adequate for more extensive synostoses or for older infants. Current surgical therapy focuses on not only treating the local synostosis but also the compensatory changes in the surrounding bone. Simple strip craniectomy fails to do this. In the younger infant, this may be sufficient due to the increased malleability of the calvarium but may require a regimen of postoperative molding. A “Pi” reconstruction is preferred for most cases of sagittal synostosis. The results are immediately recognizable and usually require no further postoperative intervention. In a series of 50 patients treated with either strip craniectomy, extended craniectomy, or total vertex craniectomy, those who had the latter procedure had a better outcome as determined by the change in the cranial

index (21). In cases of undercorrection, however, secondary surgery may be considered 12 to 18 months postoperatively.

Advancement of the fronto-orbital region in patients with coronal synostosis should restore the anterior position of the orbital bar to a level equal to the contralateral side. Excellent aesthetic outcomes have been documented with this approach (22). Either inadequate advancement or poor bony contact for healing may lead to relapse and an undesirable result. Stability of the lateral margin is aided by creation of a lateral tenon that is able to maintain contact with the surrounding calvarium after advancement. In addition, a bone block may be placed immediately posterior to the advanced tenon to prevent posterior migration.

Correction of metopic synostosis presents its own unique complications. Most authors report excellent overall results with modifications of the bilateral fronto-orbital advancement. Outcomes classified as good to excellent range from 65% to 96% (23,24). Inadequate anterior transposition of the lateral orbital rims leads to diminished prominence of the orbital bar laterally. Appropriate advancement should be attempted at the initial procedure to adequately reconstruct the entire forehead. Persistent trigonocephaly may be corrected secondarily in a number of ways. An anterior biparietal bone graft may be placed as a substitute for the similarly curved frontal bone or the orbital rim may be refractured and advanced a second time to regain prominence at the lateral margins. Further improvement may also require remodeling of the inferior midline forehead.

Lambdoid synostosis is the rarest form of synostosis and, because of its posterior position, residual deformities are probably well tolerated. True outcome data has been poorly documented.

New Deformity

Hollowing of the temporal areas may be a sequela of procedures designed to advance the orbital rim, such as in the treatment of coronal or metopic synostosis. Techniques to avoid this problem, including placement of bone fragments behind the muscle and resuturing the muscle back to the temporal bone have largely been ineffective (25). Avoidance of this complication is enhanced with lateral dissection *above* the temporalis muscle when turning down the bicoronal flap. The muscle is left intact and attached to the temporal and sphenoid bones. A secondary effect is preservation of some portion of the blood supply to the temporal bone that may provide continued vascularity and minimize the risk of isolated growth inhibition. Other options for treatment once the hollowing has occurred includes an attempt to readvance the muscle and resuspend it in a more anterior position as well as filling the area with autogenous bone graft or alloplastic substitutes, such as hydroxyapatite (26).

MIDFACIAL SURGERY

Dental Injury

Avoidance of dental injury involves an understanding of the anatomy of the teeth above (in the case of maxillary surgery), or below (in the case of mandibular surgery) the level of the gingiva. The root of the canine extends the highest of all the maxillary dentition, often to the level of the piriform aperture. roots are visible with the jaws as swellings interspersed among neighboring concavities.

In the course of exposing, osteotomizing, and plating the craniofacial skeleton, injury to the adjacent teeth may occur. Carious teeth and fractures that extend into the dental pulp should be removed to prevent infection. Attempts to retain loose teeth may be accomplished by wiring the teeth to the arch bar and applying a small amount of acrylic across the labial surface to firmly hold the teeth in place. Later infection will require removal of the tooth.

The location of any osteotomy and the placement of arch bars/screws must take into account the position of the dental roots above (or below) the gingiva. For this reason, osteotomy at the Le Fort I level should be made at least 5 mm above the estimated apex of the dental root. While the nerve supply to the teeth may be interrupted, the vascular supply preserves viability of the teeth. Screws should similarly be placed above (maxilla), below (mandible), or between

the roots if necessary. In children, most of the maxilla and mandible are occupied by developing dentition. The unerupted canines lie very close to the inferior border of the mandible. In such cases, intermaxillary fixation alone is preferred.

It is also important to be aware of the cant of the teeth. The incisors lie in the midline with minimal obliquity, whereas more posterior molars are usually canted off their vertical axis. A midline split of the palate will be less likely to injure dental roots if performed closer to the midline than more posteriorly around the molars and premolars.

Nerve Injury

The infraorbital nerve travels along the floor of the orbit and exits onto the anterior maxilla, approximately 1 cm below the orbital rim in the mid-pupillary line. Injury is best avoided by first dissecting the anterior maxilla medially along the medial buttress to the infraorbital rim and laterally along the lateral buttress before proceeding with the intervening portions. A finger should gently rest on the infraorbital rim to prevent accidental injury to the globe from dissecting too far superiorly.

Relapse

Relapse following maxillary or midfacial advancement is related to the amount of advancement, the elasticity of the overlying soft tissues, and the adequacy of fixation. An advancement of 10 to 12 mm is possible in non-cleft patients with adequate mobilization of the separated segment. In cleft cases or those with larger amounts of advancement and a tighter soft tissue envelope, the risk of relapse is higher (27). In such cases fixation with resorbable plates and screws may be less desirable. Once established, repeat osteotomy and fixation may be indicated to restore proper postoperative occlusal relationships. Distraction of the midfacial segments should be considered if relapse is a concern.

Enophthalmos

Enophthalmos results from a discrepancy between the contents and the volume of the orbit. Following trauma, some of the orbital contents may escape through fracture lines while others may atrophy. Avoidance of posttraumatic enophthalmos involves careful reduction of the fracture and replacement of the orbital contents to their original location (28). Since the malar complex articulates at four points, adequate repair requires proper reduction and fixation at three of the four. The easiest to approach are the frontozygomatic suture, the infraorbital rim, and the lateral buttress. Accurate reduction may be gauged by examining the sphenoid articulation along the lateral orbital wall (29). In the presence of a more posterior associated fracture of the zygoma, reduction at this site should be performed as well. Midfacial distraction probably results in minimal postoperative enophthalmos since no true tissue gap exists as the segments are slowly separated.

MANDIBULAR SURGERY

Nerve Injury

Avoidance of nerve injury, like dental injury, involves knowledge of the course and variety of the various nerves encountered around the mandible. The most superficial and easily injured are the marginal mandibular branch of the facial nerve and the mental nerve. The former is at risk during transcutaneous exposure of a fracture of the body or angle of the mandible. In approximately 20% of patients, branches of the nerve may lie below the inferior edge of the bone in close proximity to the facial vessels (30). The artery may be palpable at the medial aspect of the masseter muscle. The incision should lie posterior to this point and stay one to two finger-breaths below the edge of the mandible. A nerve stimulator may be also used to minimize the risk of injury during dissection.

The inferior alveolar nerve, which runs within the inferior portion of the mandible, exits through a foramen in the anterior portion of the mandible, opposite the second bicuspid.

Injury results in anesthesia of the ipsilateral lower lip, gingiva, and teeth. With osteotomies through the body of the mandible, the risk of concomitant nerve transection is rare. Despite intraoperative visualization of an intact nerve, many patients continue to display symptoms of mental nerve dysfunction months to years after the injury likely due to traction on the nerve. The mental nerve is at risk for avulsion when attempts are made to appropriately place a plate along the lower border of the mandible. Injury may be minimized by adequate exposure of the nerve and the fracture fragments. Attempts to make smaller incisions may put more traction of the nerve as visualization is limited. When recognized intraoperatively, attempts to repair the nerve under loupe magnification should be attempted.

Relapse

Relapse falls into two broad categories: immediate and late. Immediate relapse occurs with removal of interdental fixation at the conclusion of the procedure. It results from the mandible reseating itself into its proper position within the glenoid fossa and creating a class II malocclusion. Often, the mandible is plated with traction, pulling the ramus and condyle anteriorly out of the fossa, such that correction requires posterior displacement back into the fossa.

With all orthognathic cases, the procedure should not be considered complete until the interdental fixation has been removed and the jaw ranged through its normal arc of motion to assess whether or not proper occlusion with the maxilla is attained. If it is not, the plates and screws should be sequentially removed until proper occlusion, *with the mandible properly seated in the fossa*, is achieved.

Later relapse in the management of fractures is rare. In orthognathic procedures, it is more common and likely due to stretching of the overlying soft tissues seen with advancement. It has been shown to occur more commonly in patients with a higher preoperative mandibular plane angle and a more severe anterior open bite deformity (31). While some thought was given to changes in hyoid bone position as a cause of late relapse, studies have shown that hyoid position in the neck actually remains static in the postoperative period, likely due to changes in its soft tissue attachments (32).

Growth Disturbance

Growth of the mandible proceeds from the condyle inferiorly and anteriorly. Early distraction of the hypoplastic mandible, however, is indicated for the constellation of first and second branchial arch anomalies. Fractures of the mandible involving the condyle or condylar neck have caused concern in the past related to growth disturbance. This may be overstated, however, as some studies have failed to document significant growth retardation in children with high fractures of the mandible (33). Certain elective orthognathic procedures should be delayed until completion of skeletal maturity to avoid interference with lower facial growth.

GENIOPLASTY

Augmentation of the chin may be performed by insertion of alloplastic material or by osteotomy and advancement of the inferior segment. Craniofacial surgeons who are comfortable with the techniques of performing an osteotomy and fixing bone prefer osseous genioplasty. The advantages over alloplastic augmentation include avoidance of foreign material, a lower rate of infection and migration, the ability to correct severe anteroposterior deficiencies, and the absence of bone resorption beneath the implant (34,35). The only disadvantages are the use of general anesthesia, the need for saws and drills, and the potential for unsatisfactory fracture lines.

Dental Root Injury

The surgeon performing an osseous genioplasty must be cognizant of the dental root lengths of the anterior incisors and canines. Too low an osteotomy may compromise the success of the procedure; however, too high an osteotomy may injure the dental roots and lead to tooth loss.

A preoperative anteroposterior and lateral cephalogram should be obtained to identify any dental anomalies and to adequately plan the amount of advancement (or retrusion) that is ideal.

Resorption and Relapse

Resorption of bone following genioplasty may be the result of several factors. Larger-sized implants have a greater tendency towards resorption than do smaller ones. This likely relates to the tighter soft tissue envelope that is created after insertion of the implant. Resorption of bone may impinge upon the dental roots and lead to tooth loss and/or infection.

With osseous genioplasty, some resorption naturally occurs along the superior border of the advanced segment, while bone deposition occurs along the lower border in the region of the advancement (36,37). Greater degrees of resorption may be due to differences in technique. Greater amounts of muscle elevation may lead to greater degrees of bone loss following osteotomy and advancement. Earlier techniques of osseous genioplasty involved complete stripping of the musculature attachments of the chin prior to osteotomy, such that the displaced segment became a free bone graft (38). More recent techniques have advocated preserving as much muscle as possible on the chin to provide adequate vascularity to the inferior segment. This has been supported by experimental work in primates which showed pedicled segments undergo less resorption than do free graft segments (39).

Resorption must be differentiated from relapse of the displaced segment. The latter may result from several factors, including poor bony contact and/or a tight soft tissue envelope. The contact between the superior mandible and the inferior displacement segment must provide enough contact for adequate healing of bone. In the case of advancement, the preserved muscular attachments will act to pull the inferior segment back into its original position if stability through initial fixation and later osteosynthesis is not sufficient.

Advancement of bone within a limited soft tissue envelope may also contribute to relapse. This may be seen in patients with Goldenhar or Treacher Collins syndromes. In such cases, some overcorrection is probably warranted to allow for relapse due to resorption. This is not warranted in the aesthetic patient where the amount of correction should match that allowable by the laxity of the soft tissues of the chin.

Fracture

Weakness of the alveolar bone may lead to fracture of the superior portion of the mandible (Fig. 4). Treatment may involve a period of intermaxillary fixation to allow time for healing or primary fracture repair. The latter may present more of a challenge since the usual points of



FIGURE 4 Panorex following osseous genioplasty demonstrating postoperative midline fracture of the symphysis beginning at the point of fixation.

fixation along the inferior border have already been separated from the more superior segment that has fractured.

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23 | Legal Issues in Craniofacial Surgery

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INTRODUCTION

The law has a major impact on the practice of medicine subjecting physicians to duties and obligations far beyond that experienced by other professions. Imagine the press, media, or lawyers, for that matter, living under the increasingly complex system of rules and regulations that have made medicine what it is today. Lawyers make the rules, lawyers interpret them, lawyers prosecute them, and we pay lawyers to help us survive them. It is a system to which we are required to submit for the “privilege” of practicing medicine.

This chapter will cover the major areas of concern for the practicing craniofacial surgeon. Although craniofacial surgery is very special indeed, the legal issues involve the topics that trouble all physicians: medical malpractice, informed consent, and record keeping. Intentionally left out are a bewildering array of rules and regulations that issue from passage of a multitude of federal legislation, from the Occupational Safety and Health Act of 1970 (OSHA) to the Health Insurance Probability and Accountability Act of 1996 (HIPA), from the Americans with Disabilities Act of 1990 (ADA) to whatever initials they can think up next. Perhaps if an attorney spent all his time learning the latest interpretation of these laws, could he write a helpful chapter, but he would also know that it would be outdated before publication. Although there have been changes in recent years, particularly in attempts at tort reform and the impact of the Internet, principles of tort law and the standard of care have changed little and thereof are more reliably written.

As you read this chapter and experience the world of pain that is the law, think of a young Paul Tessier starting out today in America, and consider if craniofacial surgery could have ever developed in this medicolegal milieu. What wonders of medicine and surgery do we live without today and our children will live without tomorrow because of the legal times in which we live?

Regarding the references to legal cases given in this chapter, it is very helpful, educational, and occasionally enjoyable to read these cases to understand the way legal thinking is performed. It is extremely easy to find these cases using the citations given in any library having legal references. In addition, many can be found easily on the Internet. I encourage all to pursue any cases of interest.

MEDICAL MALPRACTICE

In an action for medical malpractice, the plaintiff must prove four things, each by a preponderance of the evidence (i.e., it is more likely than not). They are: (i) the physicians owed a duty to the patient; (ii) the physician’s acts or omissions did not meet the “standard of care”; (iii) this deviation from the standard of care was the proximate cause of the patient’s injury; and (iv) there was an injury that was the result of the physician’s actions or inactions. We look at each in turn, duty, standard of care, proximate causation, and damages.

Duty

For an actionable medical malpractice to occur, the physician must have owed the patient a duty of care. In the setting of a plastic surgery practice, this is almost never a point in contention. As long as a doctor–patient relationship exists, there will exist a duty. However, we

frequently ask for advice from colleagues about a particular patient. Giving such advice does not create a doctor–patient relationship between the consulted physician and the patient and therefore does not give rise to a duty. A duty may arise obviously if the patient is seen by the consulted doctor. In the emergency room setting, if one is on call and refuses to see an emergency room patient, a physician may be precluded from asserting that there was no doctor–patient relationship (1).

However, we are still free to refuse to treat a particular patient, a situation that arises frequently in plastic surgery. If one refuses to treat a patient who is part of a legally protected class *because* he is a member of that class, one will open oneself to being charged with a violation of law. For example, one could not refuse to treat a particular racial or religious group, as this would violate Civil Rights Laws. Although not covered by Federal Law, some groups are protected by some State Laws (e.g., gays). This is not a subject of malpractice law, but the issue of operating on HIV-positive patients who want cosmetic surgery is a frequent one. In the U.S. Supreme Court decision, *Bragdon v. Abbott* (2), a dentist who normally performed fillings in his office decided to treat an asymptomatic HIV-positive patient in the hospital. There was no refusal to treat, only a desire to treat differently, to perform the procedure in a hospital and avoid the possible exposure of his employees, equipment, other patients, and himself to the deadly virus. Although the dentist promised not to charge any more, the patient, facing the charges from the hospital, sued under the Americans with Disability Act. It reached the U.S. Supreme Court. Under the ADA, a disability is covered if it substantially limits a substantial major life activity. The problem here was that the patient was asymptomatic. So her side argued that she had chosen not to have children because she was HIV positive and reproduction is a major life activity; therefore, she is “disabled.” Bingo! In a 5-4 decision (Rehnquist, Thomas, Scalia, and O’Connor dissenting), the court said that an HIV-positive person was disabled from the moment of initial infection and she was covered by the ADA. These learned men and women in their clean black robes, who never have to deal with such things as blood and other bodily fluids, have decided that you may not discriminate against an HIV-positive patient. I have no doubts that this would apply to cosmetic surgery.

Standard of Care

Assuming a doctor–patient relationship with the resulting duty, there must be a violation of the standard of practice. This is one of the most confused issues in legal medicine, though the legal concept is clear. It is misunderstood by physicians frequently and intentionally misused by attorneys as well.

First, what it is not. It is not what the average doctor would do, otherwise almost 50% of physicians would not meet the standard of care. It is not what the majority of doctors would do. Most definitely, it is not what you would do! Yet, this is what one sees if one looks at some experts’ testimony. They criticize care because they would not have done that. Or they defend care because that is what they would have done. In other words, the standard of care is not determined by the majority. It is determined by some minimum standard of care that is required under the circumstances. Fall below that minimum and you have violated the standard of care. Meet the minimum and, even if the majority of physicians would not do it that way, you have not violated the standard of care. The standard is what a reasonable specialist would do in the same or similar circumstances. In the real world, this is not always explained very well to a jury.

Well accepted in jurisprudence is the “respectable minority” (3), where the law recognizes that there may be differences of opinion in the medical community regarding proper treatment. The minority opinion needs to have some acceptance in the medical community, but certainly not widespread acceptance. In plastic surgery, particularly craniofacial surgery, where treatment is so frequently tailored to the individual patient, this becomes an important principle. Attorneys unfamiliar with plastic surgery can be at a loss when trying to decide if a treatment was within the standard of care. They turn to experts to help them decide and the unknowledgeable or unscrupulous expert may start with the usual, “I would not have done that. Most plastic surgeons wouldn’t do that,” and the game begins.

A very confusing area, at least to me as an attorney and plastic surgeon, is the requirement of one using one's "best judgment." On its surface, this is a most reasonable requirement, and we surgeons cannot imagine not using our best judgment, so what is the big deal? The big deal is that this concept can be used to show a violation of the standard of care, even the care used was well beyond minimum standards of the profession. The surgeon did perform surgery within acceptable standards, but in that circumstance, he did not use his best judgment to decide to use that acceptable method or using his best judgment he should have known there were "unnecessary dangers in the community practice" (4). This concept dovetails with what standard is used. The locality rule, which meant that the care had to be up to the standards of that community, has largely been supplanted by a national standard. In the case of specialists, we are held to the standard of a specialist (even if we are not reimbursed as one). The more you are an "expert," the higher the standard of care may be. In this age of nonstop personal aggrandizement, advertising yourself as the world's greatest anything will raise the standard by which you will be held. If you are the world's greatest craniofacial surgeon, let us call you "Henry," you may be held to a very high standard indeed. Of course, in fairness, who could reasonably judge your greatness? Unfortunately, the court may allow a part-time emergency room osteopath with a substance abuse problem to testify against you.

Okay, you have met the standard of care of your subspecialty and you used your best judgment, you can breathe a sigh of relief. Not so fast! One of the most infuriating cases that a law student who happens to be a surgeon has to learn is *Helling v. Carey* (5). I read it in 1992 and I have not yet recovered. In this case, two physicians were sued because one of their patients developed blindness secondary to glaucoma during the time that the physicians were seeing the patient for routine visits. It was clearly established and accepted by the court that the physicians had not been required to test for glaucoma. They had clearly met the standard of care. The court found that the standards of the medical profession were not adequate. As the court stated, "Courts must in the end say what is required; there are precautions so imperative that even their universal disregard will not excuse their omission." In other words, when there is an innocent harmed patient and you cannot fault the doctor, you can always blame the standards that they were following. I bet we have all had a lot of glaucoma tests since 1974.

Proximate Causation

Your treatment of a patient to whom you owed a duty was below the standard of care for your profession, but did this deviation from the standard of care actually cause the injury? You used the wrong plate in a fracture and the patient developed a wound infection. The plaintiff must prove by a preponderance of the evidence that the wrong plate caused the wound infection. An expert testifies that you waited too long to do an open reduction and internal fixation of a facial fracture necessitating an osteotomy. Everything goes well, but the patient is troubled by recurrent headaches. Did the unnecessary osteotomy cause the headaches? These are questions of proximate cause. We used the term "proximate," because there has to be a reasonable causal connection between the "cause" and the "effect" (6). You are driving and you see a hamburger ad and get hungry. You stop for a bite and pulling out of the parking lot, you hit a car. The ad that you saw was a cause of the accident. If you had not seen it, you would not have pulled in for a snack, but it is not the proximate cause. The injured party cannot sue McDonalds (except in California, just kidding).

In the real world, a naïve, unknowledgeable jury is shown testimony by an expert that you violated the standard of care. You may agree, but it had nothing to do with the injury that occurred. The injury was a "mal-occurrence," something bad that happens which is not anybody's fault. The jury sees an innocent injured "victim" and a doctor who definitely violated the standard of care. The legal niceties of "proximal causation" can get lost. Your expert and attorney need to help by making this very clear to the jury.

Injury

Legally, pretty much everything is an injury. It is very difficult to argue that there is none. However, injuries are sometimes greatly exaggerated and can be attacked on that basis.

It amazes me how victims of horrible diseases manage to enjoy sex, but a plaintiff with a slightly wide scar can never have sex again. Oh, and neither can her spouse!

THE INTERNET

Marketing on the Internet is now an important component of many plastic surgeons' practices. It is very inexpensive compared with the Yellow Pages and reaches internationally. Especially for younger generations, it is where patients find out information about plastic surgery and plastic surgeons.

One must be careful about the claims made on a website. "Puffery" is accepted in commercial speech (e.g., "this is the best car ever built"), but professionals are held to a higher standard, and claims that cannot be supported by evidence are unacceptable both legally and ethically. If claims are made of special talents or abilities, the plastic surgeon may be held to this higher standard in court. In addition, it is now possible to recall on the Internet any website's appearance on a particular date. Changing your website after the fact will not erase the record of how it appeared when the patient read it.

Patients should give consent before their photographs are used on the Internet. Although this consent can be oral, it is a good practice to get it in writing. This permission should be included on your standard photographic consent. Patients, particularly those having facial surgery, may request that their photographs not be used on the Internet and this request should be honored. Failure to do so would be a breach of confidentiality as well as a breach of fiduciary duty.

Care should be used in answering e-mail generated by your website. Obviously, a written record of this communication will exist. If someone other than the physician answers the e-mail, these responses should be reviewed, since the physician will be held liable for anything said.

A complicated part of the law is where a plaintiff may be allowed to file suit. Before the Internet, you could not be subject to lawsuit in a state with which you had no connection. In legal terms, the court in that jurisdiction could not assert personal jurisdiction over you. The plaintiff had to come where you were and file suit. Jurisdictions vary in its friendliness to medical malpractice suits. In some counties, jury awards are frequent and large. This variation accounts for some of the differences in malpractice insurance premium rates. If you are in a relatively low risk state with favorable juries, you do not want to get sued in a much less favorable state. Since the Internet reaches everywhere, a physician has legally placed himself in the stream of national commerce and advertised in another state to get that patient. The patient may be able to sue you in their home state, which obviously may not only be inconvenient, but may be subject you to a very unfavorable forum. One approach to this problem is to include a "choice of law and forum" clause in your consents. This would allow patients to voluntarily agree to settle any future dispute in the courts of your state using its laws. I have no personal knowledge of any challenges or affirmations of its validity, but such clauses are used everyday in commercial contracts.

EXPERTS

Except in cases of "*res ipsa loquitur*," where "the thing speaks for itself," an expert is needed to define the standard of care and testify whether it has been violated. *Res ipsa* cases are frequently defined by statute, e.g., leaving an instrument in a patient, and no expert is needed to say that was wrong, but the usual case requires an expert. States define what the qualifications of the expert must be. With tort reform increasing in recent years, these qualifications have been tightened up, but generally, they are still fairly lax. Nevertheless, there are board-certified plastic surgeons who will testify to the most amazing things. As a member and chairman of both the Ethics Committee and Membership Committee of the American Society of Plastic Surgeons (ASPS), it has been my misfortune to read the testimony of ASPS members testifying against other ASPS members, and I can assure you that even requiring every expert to be a member of ASPS will not help much.

Defense attorneys frequently attack the opposing expert by pointing out that they always testify for the plaintiff. Consequently, defense attorneys prefer that their experts testify for both sides. Using such an expert makes him appear more credible, that he has an open mind, and believes in justice no matter which way the case falls. Hence, now that I am no longer practicing, I am willing to testify for either side depending on the merits of the case. I am not happy about testifying against a physician and I will only do it only if the negligence is clear to me. However, I know it will make me a more attractive expert to both sides, particularly to the defense. A jury expects doctors to back up other doctors. Despite the fact that we know that some plaintiffs' experts will say anything, they have immediately credibility to a jury because they are testifying against "one of their own."

An expert should obviously be well qualified and have some legal experience. However, he or she should also be able to give real help to the attorney. Defense cases in particular are frequently handled by big law firm associates who lack experience in plastic surgery cases. The surgeon being sued should educate the attorney and provide all the assistance needed, but lawsuits take such an emotional toll, this is sometimes problematic. The expert can fill in. He can also help with the deposition and testimony of the opposing expert.

MALPRACTICE INSURANCE

The cost of malpractice insurance has escalated dramatically in recent years. In some areas, it is driving physicians from that state (e.g., West Virginia). Although in the past, the thought of voluntarily going bare was unthinkable, today it is becoming an alternative. In some communities it is not possible, since hospitals may require coverage, and you may not be able to operate at outpatient surgery centers or your office without having hospital admitting privileges (e.g., Nevada). In other states, the legislature has responded by making it possible to practice bare rather than passing meaningful tort reform (e.g., Florida). I certainly do not recommend it, but I am realistic enough to know that there may be circumstances where it becomes necessary. I recommend a long talk with an attorney versed in asset protection before proceeding.

Malpractice insurance comes in two varieties, claims-made and occurrence. In the past, most policies were occurrence. Occurrence policies cover you for life for any patients you treated while you were insured. No additional coverage is required when you stop practicing to cover you when you are no longer paying premiums. Due to the long-term risks to the insurance company, these policies became prohibitively expensive or ceased all together, leaving claims-made policies to be the predominate form of insurance today. Claims-made cover you for lawsuits filed while you are paying premiums. When you no longer have claims-made insurance, you no longer have coverage for the cases that you did while you were insured. Consequently, when you move and must have another company's policy, or you retire, or go bare, you will need to have a separate policy to continue to cover you after your claims-made policy is over. This separate coverage is usually called a "tail" or "extended reporting period" coverage. Many companies will give you a tail without cost if you have been insured with them for a certain number of years and then you retire after a certain age, become disabled, or die. Everyone should check his policy to make sure this protection is present. A tail typically costs about twice your annual premium (i.e., if you are paying \$100,000 a year, a tail would cost \$200,000). Related to tail concerns is the statute of limitations.

Although tort reform has generally limited this to two years or less, courts are generous in ruling that it may not apply. Although this may be overturned on appeal, the painful process of the litigation can proceed. For plastic and craniofacial surgeons who operate on children, the statute of limitations can be very long indeed, sometimes stretching until the child is an adult, making insurance coverage all the more valuable.

A FEW THOUGHTS ON THE PROCESS

Once you have been served with a lawsuit, you have begun a painful process that is both confrontational and theatrical. Truly, all life becomes a stage. A lawsuit is a declaration of war.

Everything you say to anyone about it is discoverable and will rarely be of any value to you. Speak to your attorney and no one else without your attorney's say so. When you are to be deposed, prepare with your attorney in advance, schedule nothing else for that day, and get someone to cover your practice so there will be no distractions. Depositions are about discovery. The opposing attorney is trying to discover anything that he can use against you. Answer truthfully, but do not volunteer any information that is not necessary to answer the question. Begin every answer with a repetition of the question. When you asked your name, your answer begins with "My name is...". This gives you time to think about the answer once the questions get more complicated. Answer all questions slowly, pausing beginning your answer. You will probably want to do this when the questions get tougher, but if you have not been doing that all along, it will let the opposing attorney know when he has touched a nerve. Avoid "Yes" or "No" answers, which can paint you into a box. Put the question in your words by rephrasing it, and then answer it. Take frequent breaks that are always allowed. Never think that you can explain everything to the opposing attorney so that he can see there is no malpractice. It may simply give him more ammunition to use against you.

Attend all the depositions, particularly that of the opposing expert. Shake his hand, introduce yourself, and look at him during the entire deposition. If he can look at you more than twice, write me a letter. Your presence may affect his ability to say the more absurd things. On the other hand, when he is asked his name and he answers, "worst case of malpractice I have ever seen," your presence may not help in that way, but your attorney will feel better.

VICARIOUS LIABILITY

Vicarious liability is liability based on some legal relationship that a party has with the person who actually commits the tort. For example, if your employee has an accident while she is going to the bank for you, you are responsible in addition to the employee. Since you may be the "deep pocket," you may be the target, hence the importance of incorporating or practicing as a limited liability company instead of as a sole proprietorship. When a legal entity employs the employee, the legal entity is responsible [e.g., your professional corporation (PC)]. When you personally employ an employee, all your personal as well as business assets are at risk.

One area of vulnerability for plastic surgeons is their association with other plastic surgeons. A simple office sharing arrangement with no clearly defined legal relationship can be found to be a partnership. This kind of partnership is known as a "general partnership" and there is unlimited personal liability for each partner for the torts of any other partner or employee. One of the things that will be looked at is how the physicians present themselves to their patients. For example, if both names are on the door without any distinction between practices, if employees are shared, if the same phone number is used, the law may imply a partnership. In this case, you and/or your PC may be held completely liable for any acts of malpractice by the other physician. Basically, there must be a formal legal relationship between physicians who practice with each other to protect the other party.

Less serious, but potentially troublesome is apparent authority. Apparent authority is authority that is not actual, but appears to exist. For example, your office manager has actual authority to order supplies, schedule your surgeries, and change your Yellow Page ad. When she is fired, she no longer has any real authority. However, she is a little peeved and so orders multiple subscriptions to "Ex-Con" and "Barely Legal" magazines, cancels your ads, changes your phone number, and requests that the post office send all your mail to Sparta, Mississippi. You suffer financial damage and want to sue all these companies. Your lawyer informs you that, though she did not have actual authority, she did have apparent authority, since you had previously informed those business through your actions that she had had the authority to do those things, and you failed to notify them that she no longer had authority. Hence, from their point of view, she still had the authority, and in fact, legally she did, called apparent authority. When anyone acts as your agent, and then they no longer are your agent, notify the parties that dealt with her that she no longer has authority and you will be protected.

ASSET PROTECTION

Briefly, there are many things that you can do to protect your assets that are perfectly legal. If you should have a financial catastrophe, you will want to limit the extent of the damage. Generally, this is done by segregating assets so they are not all at risk. The best time to do this is when you can see no reason for doing it. After a lawsuit or even just after the bad outcome, it may be too late to avoid what is known as a fraudulent conveyance, a conveyance that is done for the sole purpose of avoiding paying a judgment. If you are incorporated, your personal assets are still at risk for any tort that you are found to have committed. Although a healthy malpractice insurance policy is a great comfort, judgments in excess of policy limits do occur and the surgeon should be prepared. Among asset protection attorneys, the belief is that physicians usually get into trouble with outside investments that are poorly structured, rather than with over policy limits awards.

INFORMED CONSENT

A lawsuit based solely on an alleged lack of informed consent is quite rare. More commonly, an informed consent claim is added to the main complaint of medical negligence. In the context of plastic surgery cases, the unfavorable result is frequently the basis of the suit. Whether supported by the facts or not, the patient alleges that had she been informed about the possibility of this unfavorable result, she never would have consented to the procedure.

This is used as yet another bargaining chip by the plaintiff's attorney in ongoing negotiations. Thus, obtaining and documenting the informed consent becomes a valuable exercise for the plastic surgeon.

Historically, failure to obtain permission before a medical procedure was considered the tort of battery (7). Battery is an unlawful (i.e., without permission) touching which need not be harmful in order to be actionable against the physician. Today, battery is rarely the basis of a lawsuit against a surgeon. Although a procedure without consent is an unlawful touching and no harmful result need be proved, the monetary value of damages may be quite small. In addition, battery is an intentional tort (i.e., the result of an intentional act rather than a negligent act) and will not be covered by the physician's malpractice insurance. This makes the case less attractive to a plaintiff's attorney, since a surgeon is more likely to settle a case if the settlement is paid from the insurance company's money rather than his own. Consequently, the claim will be filed as a medical negligence case rather than a battery action to bring it within the insurance policy's coverage. This approach is widely accepted by the courts, since failure to obtain consent before a procedure is considered a breach of the standard of care and, consequently, is an act of medical malpractice. Surprisingly, it was not until 1957 that the term "informed consent" was first used by a court (8). Following the 1960s, legal theories of patient autonomy expanded and the patient became a partner in the doctor-patient relationship entitled to an adequate understanding of contemplated treatment plans. Physicians have a legal and ethical obligation to obtain an informed consent from their patients prior to treatment. The duty to obtain consent before treatment is rooted, firstly, in the long recognized principle of English common law that no free man may be touched without his permission. Secondly, a right of privacy and individual autonomy that gives an individual sovereignty over his own body has been elucidated by the U.S. Supreme Court as flowing from the Bill of Rights and the 14th Amendment.

ELEMENTS REQUIRED IN AN INFORMED CONSENT

It is easy to enumerate the kinds of information required to be disclosed to the patient, but it is much more complicated task to list the details of what must be disclosed. In some cases, it will depend upon which state the surgeon practices. In other cases, it will depend on the individual nature of the particular patient. These complicating factors will be discussed later in the chapter. In general, five types of information are required and additional information may be required stemming from the physician's role as a fiduciary. The standard five types of information required are the following.

Diagnosis

This is rarely an important issue in the context of plastic surgery, and it is rarely litigated.

Nature and Purpose of the Proposed Treatment

A detailed description of the procedure is not required, and although it may be good practice, there is no case law requiring the discussion of the likelihood of success (9).

Availability of Alternative Treatments

This is an extensively litigated area. The patient must be informed about any medically acceptable alternative treatment that could be used under the circumstances. This includes procedures that the informing physician could not, in fact, perform, and the patient would have to see another physician for the alternative treatment. It is not, however, necessary to inform the patient about procedures that neither the informing physician nor another specialist would recommend (10).

Risks, Complications, and Consequences

This is clearly the “mother lode” of informed consent litigation in plastic surgery. More frequently than is justified by the facts, there is an unfavorable result, a suit for malpractice is filed, and an informed consent claim is added claiming that the patient never would have had the procedure if she had been informed of the possibility of that specific unfavorable result. A risk or complication is something that might happen as a result of the procedure, e.g., a postoperative hematoma occurs after a facelift. A consequence is something that either normally occurs after a procedure, e.g., a joint will not move after an arthrodesis, or is something that results from the occurrence of a complication, e.g., skin loss following a post-facelift hematoma.

Courts do not require that every risk be disclosed to a patient. Extremely unlikely risks are generally not required to be disclosed, but this is subject to exceptions in some states. Case law indicates that risks that are “commonly known” need not be discussed (11), but in our present medicolegal milieu, reliance on this by the physician is not recommended to avoid being second-guessed later. Finally, although rarely mentioned in talks and in the literature directed at physicians, what you legally need to disclose to a patient is dependent on the informed consent standard adopted in your particular state which will be addressed shortly.

Result if No Treatment

A physician is under the legal duty to inform the patient about any unfavorable results that may occur without treatment. For example, a biopsy of a suspicious lesion may be indicated, but the patient refuses. The patient must be informed that the result of not having a biopsy may be a delay in diagnosis of a malignancy increasing the extent of later treatment or raising the probability of mortality (12).

THE PHYSICIAN AS FIDUCIARY

The physician–patient relationship imposes on the physician the highest standard of duty imposed by law, that of the fiduciary. A fiduciary is one who undertakes to act in the interest of another person while subordinating the fiduciary’s personal interests to that of the other person (13). This has led a court to find that a physician had a duty to disclose his alcoholism to a patient (5,14) and another to find a duty to disclose a physician’s HIV-positive status (15).

Although not strictly an issue of informed consent, physicians are obligated to disclose a failure of treatment or the happening of a mal-occurrence during a procedure (e.g., “the nerve was accidentally cut”) (16).

STATE VARIATIONS IN DUTY TO DISCLOSE

There are two standards used to judge the adequacy of an informed consent, the physician-based standard (17) and the patient-based standard (18). Some states use a hybrid rule (19). Although the number of physician-based states (at 22) outnumber the patient-based states (20 plus DC), a majority of the hybrid states are more patient oriented than physician oriented, effectively making the patient-based standard the majority rule in the United States.

Initially, courts allowed the medical profession to set the standards for informed consent. What was required was what the doctors in the community thought was necessary. This evolved into the physician-based standard. Within the past few decades, some states have, instead, adopted a rule that requires disclosure of what the patient would consider material to his decision. This is the patient-based standard.

Physician-Based Standard

A plastic surgeon in a physician-based standard state needs to inform the patient what the reasonable plastic surgeon would tell the patient under the same or similar circumstances (20). This standard is basically the same as that used for medical malpractice where the question is whether the physician violated the standard of care as determined by the physician's specialty group.

Such a standard can only be determined by an expert witness who can testify to the standards of the specialty. In an informed consent case in a physician-based standard state, the prevailing standard of informed consent in that specialty must be testified to by an expert witness.

Patient-Based Standard

In a patient-based standard state, the plastic surgeon must give the patient all the information "material" to the decision that the patient must make rather than inform the patient about what plastic surgeons think the patient should know, the information given must be what the patient needs to know to make a decision. What the patient needs to know is what a patient would find important (i.e., material) in making the decision. Complicating matters is whether you need to tell that patient what that particular patient needs to know (subjective standard) or is it adequate to tell the patient what the reasonable and prudent patient would want to know under the same or similar circumstances (objective standard). In the groundbreaking case that first elucidated the patient-based standard (21), the court applied the "objective" patient-based standard. By far, the objective standard is the most commonly adopted standard in the states utilizing the patient-based approach. However, even using the objective approach, where you must make the patient aware of risks that the theoretical reasonable patient would want to know, the courts still insist that a physician take into account any special fears, values or sensibilities of his particular patient. The disadvantage for the physician-defendant in a patient-based standard state is that the plaintiff-patient need not have an expert witness testify that the physician should have told him before surgery about a particular risk. Since the standard is what the reasonable patient-layman would want to know, the jury can simply put itself in the reasonable patient's position and decide whether the members of the jury themselves would have wanted to be informed. From the plaintiff attorney's point of view, this is much better position to be in, because, almost regardless of the weakness of his case, he is not dependent on finding an expert and the judge will most likely allow the case to go to the jury to let the jury decide the issue. Juries, unfortunately, do not always decide rationally.

Causation Test

Even if the patient can convince the jury that she should have been told, the case is not over. The patient must also prove causation. First, she must show that the procedure done actually caused her injury, and that the physician should have known that this injury could occur following the procedure. This is usually straightforward, but an expert medical witness is usually necessary. Second, she must show that had she been informed of the risk, she would not

have had the procedure. For example, a patient has a complete facial nerve palsy after a deep plane facelift. She contends that had she known that a nerve injury was possible, she would not have had the procedure. This is possible. Another patient has a squamous cell carcinoma and develops a postoperative infection. He contends that he would have never had his cancer removed, had he been told that he could develop a wound infection. This is not believable. In the latter case, the causality test fails because there is no cause and effect relationship between not being told about the complication and having the procedure in the first place. The jury needs to decide whether the patient would have had the procedure, had he been told. States then use yet another objective or subjective standard. The great majority of states use the objective decision causality test. They ask if the reasonable and prudent patient would have not had the procedure had they been told about the risk in question. In a minority of states, the subjective decision causality test is used, and the jury tries to decide whether this particular patient would have had the procedure or not.

Hybrid Rule States

These states have generally modified either the physician-based or patient-based rules by statute. Texas, for example, created a Medical Disclosure Panel to prescribe what needed to be disclosed for particular procedures of their choosing. However, the courts found that this statute had the effect of replacing the physician-based standard with the patient-based standard for any procedures that the panel did not address (22). In Florida, the disclosure must meet the physician-based standard and be sufficient for a reasonable person to have a general understanding of the procedure, medically acceptable alternatives, and substantial risks inherent in the procedure (23). Some states have statutes covering specific situations, e.g., what must be disclosed in the treatment of breast cancer (e.g., California, Maine, Massachusetts, Minnesota).

WHEN AN INFORMED CONSENT IS NOT REQUIRED

There are a number of situations when an informed consent is not required.

Waiver

A competent patient may waive an informed consent. Patients have the right to refuse disclosures otherwise required for an informed consent and may not be forced to hear them. A waiver must be voluntarily given without duress, and the patient must be aware that they have the right to hear the information.

It is suggested that a physician give the patient enough information so that it is clear what types of information of which the patient is waiving the disclosure. Physicians are urged to document the patient's waiver.

Emergency Treatment

When prompt treatment is needed and the patient is incapable of giving an informed consent, courts generally will allow consent by proxy from the patient's nearest available relation. If no such proxy is available or known, the courts presume that the patient would want such emergency care if he were able to consent absent evidence to the contrary (e.g., "Living Will"). However, states differ on how serious the emergency need be to allow the physician a free hand. Certainly, the physician may proceed if necessary to preserve life and limb. If consent is refused by the next of kin, the physician is put in a difficult situation. If time is available, emergency permission from the court may be obtained. Hospital legal departments are usually very helpful in such situations. If there is no time, the surgeon is again put in a difficult position. If treatment is refused by a spouse and the surgeon does not proceed and the patient loses a limb, legally he should be on strong ground, but allegations of inadequately informing the relative may later come up. In this situation where a relative refuses needed treatment, it is advisable to get another party involved to stress the importance of action. If the surgeon goes ahead with treatment despite a relative's objection and in the absence of court permission, the surgeon opens himself up not only to a failure to obtain an informed consent (medical

malpractice), but also to a battery charge if permitted by the jurisdiction. Battery, an intentional tort, will probably be excluded from coverage by his malpractice policy and his personal assets will be at risk. This gives the plaintiff a very strong bargaining position.

Therapeutic Privilege

A physician may abstain from a complete disclosure to the patient when, in the physician's sound judgment, the disclosure itself poses a significant risk of harming the patient. For example, a physician decided that a seriously ill patient with hypertension and a suspected thoracic aortic aneurysm should not be told the risks of thoracic aortography. The patient developed paralysis after the aortogram and contended that had he been told of the risk, he would not have consented. The court held for the doctor on the basis of therapeutic privilege (24).

Such situations should be rare in plastic surgery, and although not required by law, it has been suggested by courts that the surgeon disclose the risks to a spouse or other close relation despite the fact that this may be a breach of confidentiality. Documentation of the reasons for the physician's action should be noted in the patient's record. In addition, a second concurring opinion by an uninvolved physician would be helpful should legal complications later occur. Fear that the patient will not have the procedure if informed of the risk is not a situation where the therapeutic privilege will apply. The constitutional right to refuse treatment even where strongly indicated is the basis of the right to an informed consent.

Patient's Prior Knowledge

Where the physician is aware that the patient has adequate prior knowledge, it is not necessary to inform the patient again. Also, courts have repeatedly stated that it is not necessary to inform patients about matters within common knowledge or about risks that are very remote. However, even if the risk is remote, if it involves death, paralysis, loss of sexual or reproductive function, or other very serious unfavorable result, it might be required. Thus, it is a safer practice to mention it.

Implied Consent

A patient's conduct may legally imply consent. For example, a patient poses for preoperative photographs in the office without objection has impliedly consented to be photographed.

SPECIAL PATIENT CATEGORIES

Minors

By statute, adulthood begins at the age of 18 years in most states (19 years in Alabama, Nebraska, Wyoming; 21 years in Mississippi and Missouri) (25). Although it is commonly believed that only the parent's consent is needed to treat an adolescent minor, this is not generally correct. Many states have passed statutes that allow minors varying from the age of 14 to 16 years to consent to any form of medical treatment. In addition, many states have adopted the "mature minor" rule that does not specify an age, but rather allows minors who understand and appreciate the consequences of the proposed treatment to give consent (26). This has a great deal of relevance to plastic surgery. A 16-year-old rhinoplasty patient must herself give an informed consent to the procedure. Generally, the more unnecessary the procedure (e.g., cosmetic), the more important is the minor's consent. Interestingly, there has not been a recorded case in the last 30 years in which a parent has recovered damages from a physician for appropriate treatment for which consent had not been given by the parent but was given by the minor patient who was over the age of 15 years (27). In the situation where the failure to treat would result in serious injury, the physician may petition the court for permission to treat in the absence of parental or minor consent.

When parents are separated or divorced, the parent with legal custody or actual possession of the child can give consent. Where the child is with the noncustodial parent

and needs treatment for an injury, the noncustodial parent may also give consent. In the emergency setting, courts are very willing to let the physician proceed with treatment when a parent cannot be reached for consent even in the absence of a serious injury. Courts do not want children in pain or frightened for prolonged periods while waiting for their parents to be found.

Incompetent Patients

When the patient has been legally adjudicated as incompetent, consent is obtained from the individual designated as guardian by the court. Usually, however, the circumstances are not that convenient. If a patient does not give consent to a needed procedure because he does not appear to be mentally competent to understand the seriousness of the situation, there is no easy answer for the surgeon. The law presumes that an adult is competent unless adjudicated otherwise. Short of petitioning the court for a guardian to be appointed, surgeons should try to enlist the support of the patient's family, ask for a second medical opinion, and discuss the situation with the hospital's attorney.

WHEN THE PROCEDURE EXCEEDS THE CONSENT

The Extension Doctrine

The extension doctrine allows the surgeon to extend the scope of the procedure when unforeseen circumstances arise, which make it advisable to do so.

This doctrine does not allow a surgeon *carte blanche*. If the extension was foreseeable and the patient was not informed about its possibility, then the doctrine does not apply. Similarly, it will not apply in situations where the extension is not so urgent that it cannot be postponed until the patient can give consent. For example, during an authorized oophorectomy, a surgeon removed a suspicious mole from a patient's thigh without consent. The court held against the surgeon, since there was no necessity to proceed without first getting the patient's consent (28). In another case, a surgeon harvested tensor fascia lata without consent to use in an authorized hand procedure. The court found that surgeon was unjustified (29). The additional procedure was a foreseeable possibility about which the patient should have been informed.

Unauthorized Surgeon

A substitute surgeon may not be used without the patient's consent. In an emergency, necessary treatment may be instituted by an unauthorized physician.

Documentation Contrary to common belief, a consent need not be in writing. What a written consent may provide, however, is documentation that the informed consent actually took place. This frequently becomes an issue in litigation following an unfavorable result. Strictly speaking, a verbal disclosure meeting the legal requirements followed by a verbal assent by the patient is all that is required. However, the surgeon, for his own protection, should record in the patient's medical record that an informed consent was given by the patient.

Listing the risks discussed may also be helpful. Another approach is to have a written consent to assure that the legal requirements are fulfilled. A very complete written consent signed by the patient is not a complete defense. Plaintiffs may contend that they were not given enough time to read it, that they did not understand it, and had no opportunity to ask questions, in short that they did not know what they were doing. There is no foolproof way of preventing an informed consent claim, but some states, bowing to tort reform, have helped to make a defense easier if certain conditions are met. At least 12 states (Florida, Georgia, Idaho, Iowa, Los Angeles, Maine, Nevada, North Carolina, Oregon, Texas, Utah, and Washington) make a signed consent form presumptively valid (30). The list of required disclosures is generally short. Plastic surgeons practicing in these states should consider taking advantage of their states' laws and use a written consent. Courts have generally looked unfavorably at "blanket" consent forms that give the surgeon virtually unlimited discretion in performing the procedure.

Courts will strike down consents that include waivers of prospective liability as void against public policy. Such waivers to sue physicians for future possible malpractice are never valid.

MEDICAL RECORDS

Physicians have a legal and ethical obligation to maintain adequate medical records. The importance of the patient's medical record in our litigious society cannot be overstated. Remarkably, medical evidence is thought to be involved in three-fourths of all civil cases and about one-fourth of criminal cases brought to trial (31). The medical record is vital in the communication of medical information among physicians and other healthcare entities. It allows continuity of care when the patient changes physicians allowing future medical professionals to evaluate the patient with the benefit of knowing what came before. More narrowly, the medical record allows the physician to clearly document the progress of the patient's treatment. This will be of major importance should a legal dispute arise in the future.

Medical records should be accurate and timely. A note should reflect each patient visit. Although very difficult to be consistently carried out, it is a good practice to make notations for every patient contact including telephone calls. The patient's noncompliance with treatment plans and failure to return for office visits should be documented. Patient's complaints and dissatisfactions should be noted. When dealing with pediatric patients, it is the parent or guardian who will determine compliance with treatment and discussions with them should be noted. Documentation of informed consents should be reflected in the chart.

The record should be legible. Inadequate medical records could be the basis of the court finding a violation of the standard of care. There are state and federal statutes which mandate that medical records be retained for a specific number of years, usually 5 to 15 years and longer for children (32). If a patient decides to see another physician, a copy of the medical record should be sent to the new physician when requested. In plastic surgery, copies of the patient's photographs are particularly useful to the new surgeon. If a surgeon moves or retires from practice, his records still need to be retained at least for the statutorily mandated time. Ideally, they could be transferred to the surgeon taking over the practice. Otherwise, there still needs to be some way for the patients to gain access to them.

Alteration of Medical Records Errors in medical records are frequent and usually involve a spelling or transcription error. However, an error as simple as transcribing "there is evidence of necrosis" when what was dictated was "there is no evidence of necrosis" could make a great deal of difference in a malpractice suit for a delay in the treatment of a complication. In addition, simple but costly errors such as these are difficult to pick up without carefully reading the record. Mistakes should be corrected as soon as possible. The proper way to correct a medical record is to put a simple line through the improper entry, so that it still can be read. Then, the appropriate entry should be placed nearby with the date of the correction added followed by the corrector's initials, which in most cases would be the physician who dictated the note. If it is not obvious from the context why the change needed to be made, then the reason should be noted. Never try to erase or obliterate an error. Never substitute an entirely new note for the old one with the intention of making the evidence of the error disappear. When the worst happens and your own words, whether an erroneous entry or not, will damn you in a malpractice case, never, never, never attempt to cover it up. You must always assume that the opposing party already has a copy of the record and would love to find an altered record on discovery. In some states, it is a criminal offense to falsify a medical record to conceal negligence or a criminal act (33). The physician would be looking at the possible loss of his medical license and a term in the penitentiary. Physicians should know that the science of documentation examination has advanced to such a point that it is very difficult to successfully falsify a medical record. Plaintiff attorneys may use such experts for suspicious entries, since the effects of finding such an alteration mean a virtually guaranteed win. As an example, did you know that the ink used in pens is labeled for the year in which it manufactured? Try explaining a 1997

entry with 1999 ink! The greatest temptation to alter a record is when the physician is really blameless and the entry was an honest mistake. Never attempt to alter the record in any way except the correct way. In this situation, discuss this with your attorney before making a change in a case which is or will be involved in litigation.

CONFIDENTIALITY

Flowing from the fiduciary physician–patient relationship is the duty to preserve the patient’s privacy and keep his medical records confidential. Legally, the physical medical record belongs to the physician, but the information contained within belongs to the patient. State laws vary, but the patient largely has a right to a copy of his records. Exceptions usually exist for psychiatric records. Unless there is a statute providing an exception, the physician may not release the patient’s medical records without the patient permission. Even a request from the patient’s attorney or the patient’s insurance company cannot be released without the patient’s consent. A good office policy is not to allow any medical record to leave the office without it being checked by the physician. Filing errors occur and it is possible to send someone else’s note or laboratory result that was misfiled into another patient’s chart. Prior to sending the copy of the record out, the presence of the patient’s consent can be verified. Such consent should be in writing.

All information about the patient is confidential. It is the patient’s privilege to waive, and with rare legal exception, the physician may not reveal the information about the patient without permission. For example, in one case a plastic surgeon used a patient’s pre- and postoperative photographs in a presentation at a department store and on television without permission, the court found that this was a violation of the patient’s right to confidentiality (34).

Breach of confidentiality can lead to a lawsuit on multiple grounds, e.g., breach of privacy, breach of confidentiality, breach of fiduciary duty, breach of loyalty, breach of contract, negligence, infliction of emotional distress, as well as liability for violation of privacy statutes (35). By statute, the patient usually waives his right to confidentiality when he puts his health in question in litigation, e.g., a malpractice action.

A FINAL WORD

The devil visited a lawyer’s office and made him an offer. “I can arrange some things for you,” the devil said. “I’ll increase your income five-fold. Your partners will love you; your clients will respect you; you’ll have four months of vacation each year and live to be a hundred. All I require in return is that your wife’s soul, your childrens’ souls, and their childrens’ souls rot in hell for eternity.” The lawyer thought for a moment and asked, “So, what’s the catch?” (36).

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24 Research Directions in Craniofacial Surgery

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INTRODUCTION

The annual growth in U.S. healthcare expenditure has dramatically increased since the turn of the millennium (1). The biomedical burden imparted by surgery on the craniofacial skeleton alone has accounted for over \$585 million in medical care, with over 16,000 craniotomies/craniectomies and 32,000 posttraumatic facial reconstructions performed in 2002 (2). Considering the unabated demand and burgeoning costs, craniofacial research has endeavored to develop new, more effective approaches to treat the multitude of conditions currently faced by clinicians. Three main areas of investigation have emerged as key elements for continued advancement in the field of plastic surgery: craniosynostosis and its underlying developmental and molecular underpinnings, the mechanics and biomolecular cascades involved in distraction osteogenesis, and the definition of cellular building blocks and bioengineered scaffolds for bone tissue engineering. This chapter will specifically focus on these areas and present recent findings which will continue to help navigate investigations over the next decade. Through insights gained into these guiding principles, it is hoped that novel treatment strategies may be developed which may ultimately translate into improved clinical outcomes.

CRANIOSYNOSTOSIS

Normal calvarial development is contingent upon coordinated growth between the brain and overlying skeletal elements. With the brain normally doubling its volume within the first two years of life, much of this increase is facilitated by fibrous joints interposed between adjacent bones of the skull vault. Natural allometric expansion of the calvarial vault is dependent on complex morphogenetic mechanisms coupling interactions between the brain, dura mater, suture mesenchyme, and bone plates. Minor perturbations of such interactions may therefore result in pathologic development. Craniosynostosis, the premature fusion of one or more sutures, restricts the expanding brain by preventing growth at the sutures. Such constraint may result in significant morphologic deformities of the craniofacial skeleton secondary to compensatory growth in regions less restricted (3). Cranial abnormalities due to premature pathologic fusion have been reported with a worldwide incidence of 1:2000 live births (4,5). While craniosynostosis has been associated with multiple syndromes, the vast majority of cases remain nonsyndromic with a sporadic pattern of occurrence. Aside from obvious phenotypic craniofacial deformities, several serious functional aspects have also been implicated with premature suture fusion including elevated intracranial pressure, seizures, papilledema, optic nerve atrophy/blindness, and hydrocephalus (6). Undoubtedly, the premature fusion of cranial sutures represents a potential harbinger of significant medical comorbidities.

Attempts at treatment and prevention of such complications have driven the considerable evolution of surgical therapy since the late 19th century. Children with pathologic fusion, however, still face complex and challenging procedures aimed at increasing cranial volume while remodeling both the skull vault and facial deformities. Such procedures have a reported mortality rate in the range of 1.5% to 2% (7). In addition, the potential for refusion in the surgically remodeled skull remains ever present, mitigating the volumetric gains made in the operating room and predisposing patients to unplanned additional surgical procedures. Considering such potential for morbidity and mortality, and given the high incidence of suture fusion worldwide, craniosynostosis clearly represents a significant medical burden (8). While recent advances in developmental biology and genetics have elucidated some of the events governing suture fate, much of the molecular signaling remains ill defined. Knowledge and comprehension of the interactions between suture components may therefore facilitate improved treatment of craniosynostosis and the prevention of its sequelae.

Cranial Development

To better understand the development and underlying mechanisms guiding cranial suture fate, investigators have primarily turned to the murine model. In both rats and mice, the posterior frontal (PF) suture fuses in a predictable manner, whereas both the sagittal (SAG) and coronal (COR) sutures remain patent. This differential suture fate has been exploited by researchers as a model to examine the cellular and molecular events during suture closure. As clinical samples represent only a "snapshot" for examination of postpathologic fusion, the incorporation of studies using rats and mice have enabled a more detailed definition of the developmental and biomolecular events occurring before, during, and after suture fusion. In addition, the high degree of genetic homology between humans and mice, including conservation of signaling pathways, has allowed researchers to gain further comprehension and to potentially develop avenues of therapy which may ultimately be translated into clinical practice.

Employing the resources available in transgenic mice, a more thorough understanding of calvarial bone and suture development has emerged, providing a novel interpretation for the embryologic origins of the skull vault. Prior work using the quail-chick chimera model had yielded contradictory findings, with bones of the cranium generally thought of as derivatives of embryologic neural crest cells (9–13). Recent studies by Jiang and colleagues, however, have contributed novel insight into these tissue origins, challenging results of previous studies (14). Using a transgenic mouse expressing *lacZ* under a neural-crest specific *Wnt1* promoter, tissues of neural crest origin could be indelibly marked and demonstrated through histologic staining (14). This model enabled detailed determination of embryologic tissue derivation for calvarial bones and their associated sutures. Interestingly, while the dura mater and frontal bones were entirely of neural crest origin, parietal bones were observed to arise from paraxial mesoderm (14). Furthermore, both the patent SAG and COR sutures were noted to form at the interface between neural crest and mesoderm tissues (SAG suture mesenchyme is neural crest derived) while the fusing PF suture was found to be comprised entirely of neural crest (14). These findings raise the potential for tissue-specific interactions which could conceivably play a major role in determining fusion or patency. The interplay between tissues of neural crest and mesodermal origin may thus dictate normal patterning and ultimate suture fate.

Molecular Biology

Several studies have been performed examining the roles of growth factors and cytokines governing suture fate. The significance of fibroblast growth factor (FGF) signaling in suture development has long been established, as mutations in their receptors have been associated with several forms of human craniosynostosis syndromes (15–20). Of the four known FGF receptors, mutations in three have been linked to premature pathologic fusion. Given these findings, significant work has been performed to delineate both the function of these receptors and their ligands with respect to suture development. FGF receptor mutations associated with human craniosynostosis syndromes have been well characterized, as several different amino acid substitutions have been implicated in receptor gain-of-function. Such mutations localized

to the IgII–IgIII linker region have been most closely associated with premature suture fusion in humans. Work by Anderson and Wilkie et al. have purported that these amino acid substitutions between the IgII and IgIII extracellular domains enhance receptor affinity for FGF ligand, thus favoring receptor dimerization and intensifying signal transduction (21,22). Furthermore, recent x-ray crystallographic analysis has demonstrated additional intermolecular contacts between mutant FGF-receptors and their ligands, suggesting a structural explanation for receptor gain-of-function (23,24). By stabilizing FGF and FGF receptor interactions, mutations in the IgII–IgIII linker region therefore serve to further promote receptor activation and upregulation of the intracellular signaling pathway.

As these studies have clearly implicated gain-of-function FGF receptor mutations in the pathogenesis of human craniosynostosis, extensive research has endeavored to define the downstream biochemical events resulting in enhanced osteogenesis and ultimate suture fusion. Investigations by Moore and colleagues have demonstrated that reduction in FGF2 signaling, through introduction of FGF2 antibody coated beads, limits both osteoblast proliferation and differentiation (25). In contrast, application of rhFGF2 soaked beads onto the calvaria of E15 mice was shown to induce ectopic osteopontin and osteonectin expression, markers of osteoblast differentiation, with concomitant decrease in cellular proliferation (26,27). These data provide a potential biomolecular explanation for increased bone formation observed and subsequent suture fusion in response to enhanced FGF signaling.

Extending these findings, transgenic mice have recently been created with similar FGF receptor mutations to those noted in human craniosynostosis syndromes. Analogous to the fibroblast growth factor receptor 1 (FGFR1) Pro252Arg mutation in Pfeiffer syndrome, Zhou and colleagues have reported a mouse line carrying the FGFR1 Pro250Arg mutation to exhibit pathologic SAG and COR suture fusion, facial asymmetry, and midface hypoplasia (28). In addition, precocious PF suture fusion has also been reported in these mice (29). In vitro studies on osteoblasts with this mutation have demonstrated accelerated capacity for extracellular mineralization accompanied by elevated expression of osteopontin, osteocalcin, and bone sialoprotein (28). Such findings suggest that gain-of-function mutations in FGFR1 may thus precipitate premature suture fusion through promotion of osteoblast differentiation and bone deposition. Parallel studies have also been performed on the fibroblast growth factor receptor 2 (FGFR2) mutation associated with Apert syndrome; mice carrying the corresponding Ser250Trp substitution have brachycephalic skulls secondary to premature COR suture fusion (30). Interestingly, while the FGFR1 Pro250Arg mutation has been associated with increased osteoblast differentiation, in vitro studies on osteoblasts carrying the Ser250Trp FGFR2 mutation has instead shown increased bax expression, suggesting that programmed cell death could potentially mediate the pathologic fusion observed (30). These findings thus raise the possibility that varying biomolecular mechanisms may be employed by different gain-of-function FGF receptor mutations toward a similar end of craniosynostosis.

In contrast to these investigations, other studies have also looked to abrogate FGF signaling through the elimination of receptor expression. Research with transgenic mice carrying a conditional knockout of the FGFR2 locus have revealed a distinct dwarfism that develops early in the postnatal period (31). Multiple skeletal abnormalities were also noted, with decreased bone mineral density relative to age-matched controls (31,32). Investigation of the calvarium demonstrated abnormal patency of the PF suture and defective osteogenesis. Work by our own laboratory has provided corollary findings for FGFR1 through use of a truncated, dominant-negative form of this receptor. In vitro transfection of this gene into the dura mater beneath the PF suture eliminated expected suture fusion on histologic analysis (33). This was confirmed through in vivo adenoviral-mediated delivery of the dominant-negative FGFR1 construct to the PF suture-associated dura mater in embryonic rats; widely patent PF sutures were observed when analyzed at 30 days of life. Clearly, FGFs and their receptors play central roles in the determination of suture fate. Elimination of FGF receptor expression has yielded a wealth of insight into the downstream biochemical pathways involved in suture fusion. With new techniques—e.g., RNA interference—to study abrogation of FGF receptors now available, use of these tools may assist in future analyses of FGF transduction pathways and in the elucidation of how increased FGF signaling ultimately contributes to pathologic suture fusion.

Though several of these investigations have endeavored to characterize the biology of FGF receptor signaling, recent studies have linked *Msx2* expression with FGF activity. *Msx2*, a homeobox-containing gene, has been shown to be associated with Boston type craniosynostosis. Using heparin acrylic beads to deliver FGF ligands to mouse calvaria, Ignelzi et al. demonstrated increased *Msx2* gene expression along with *Runx2/Cbfa1*, bone sialoprotein, and osteocalcin (34). Examination of calvarial sutures revealed pathologic COR suture obliteration where the greatest increase in *Msx2* expression was also noted (34). Extending these findings, transgenic mice carrying analogous gain-of-function *Msx2* mutations to that found in humans were noted to show enhanced parietal bone growth and pathologic SAG suture fusion (35,36). Interestingly, studies in humans with congenital enlarged parietal foramina have revealed a potential dose-dependent effect for *Msx2* in bone development. Functional haploinsufficiency of *Msx2* through heterozygous mutations has been noted in several families demonstrating calvarial ossification defects (37,38). Furthermore, investigations localizing the expression of *Msx2* in mouse calvariae have revealed a potential temporal and spatial function in suture morphogenesis (35). While *Msx2* transcripts were abundantly found in the suture mesenchyme of embryonic mice, neonatal expression was limited to osteogenic fronts (35,39). These data therefore suggest that *Msx2* may be a downstream target of FGF signaling and is differentially expressed during suture development. Craniosynostosis through upregulation of FGF activity may thus be mediated through enhanced *Msx2* transcription effecting changes in osteogenesis and suture fate.

Similar to *Msx2*, *Twist* has recently emerged as a gene potentially involved in the direction of cellular response secondary to the FGF signaling. Originally identified in embryonic gastrulation and mesoderm formation, *Twist* is a basic helix-loop-helix transcription factor capable of binding E-box regions on DNA to activate and/or repress transcription (40,41). In humans, heterozygous mutations or deletions in this gene have been associated with Saethre-Chotzen syndrome, an autosomal dominant craniosynostotic syndrome (42,43). Similarly, mice heterozygous for *twist* demonstrate many of the abnormalities observed in their human counterparts, with partial or complete COR suture fusion (44). Experiments using human osteoblasts have demonstrated downregulation of *Twist* through antisense transfection to result in a more highly differentiated state, as evidenced by increased alkaline phosphatase, collagen I, and osteopontin expression (45). In contrast, overexpression of *Twist* led to a more undifferentiated, spindle-shaped phenotype and suppression of both alkaline phosphatase and collagen I transcripts (45). Surprisingly, overexpression of *Twist* also inhibited FGF-2 downstream effects as determined by inhibition of early growth response element-1, a known mediator of FGF signaling (45,46). Therefore, *Twist* may be critical for the maintenance of a slowly dividing osteoprogenitor state, allowing for continued suture patency. In the absence of *Twist*, premature osteoblast differentiation may result with subsequent pathologic fusion.

Like *Msx2* and *Twist*, the bone morphogenetic protein (BMP) antagonist *Noggin* has become another protein closely associated with FGF signaling and a potentially significant factor in the determination of suture fate. Originally described in *Xenopus* by Smith and Harland, *noggin* was noted to play a role in normal embryonic dorsal-ventral patterning (47). More recently, *Noggin* has been demonstrated to directly antagonize BMP activity, with a dose-dependent inhibition of rhBMP4 effects on bone marrow stromal cells (48). Crystallographic analysis of *Noggin* bound to BMP7 has revealed a conformational shift in structure for BMP7, resulting in the masking of epitopes for BMP receptors type I and type II (49). With regard to suture development, *Noggin* has increasingly emerged as an important mediator of suture patency critically tied to FGF activity. Studies by Warren and colleagues have revealed *noggin* expression to be primarily limited to both the patent SAG and COR suture in mice (50). Furthermore, increased FGF2 activity led to *noggin* suppression in vitro and pathologic COR suture fusion in vivo. In contrast, overexpression of *noggin* in the PF suture of mice resulted in abnormal widely patent sutures (50). Complementary studies examining effects of *noggin* abrogation in the SAG and COR suture, however, have been limited by embryonic lethality in *noggin* deficient mice; *noggin* null mice exhibit multiple vertebral, rib, and limb defects (51). Novel approaches to the elimination of *noggin*, through RNA interference or conditional knock-outs, may assist in further defining the role *noggin* plays in suture development.

Investigations using these models may ultimately help to elucidate the interplay between Noggin, BMPs, and FGF signaling in determination of suture fate.

Other BMP antagonists, aside from Noggin, have also recently emerged, with potential roles in mediating suture development. Microarray analysis of rat cranial sutures revealed BMP3 to possess an expression pattern highly suggestive of an osteogenic antagonist (52,53). Originally purified from bone, rhBMP3 was found to inhibit embryonic ventralization, implying a BMP antagonist function similar to that of Noggin (54). In addition, rhBMP3 was found to inhibit rhBMP-2 mediated differentiation of C2C12 cells along an osteoblastic lineage (55). Unlike Noggin, however, BMP3 does not bind to other BMP ligands. Rather, BMP3 instead mediates its effects through Activin receptors and competition for shared Smad proteins (55,56). Gene analysis of BMP3 in rat calvarial sutures revealed increased expression in the SAG suture relative to the PF suture during periods of expected fusion (53). BMP3 thus demonstrates a temporospatial pattern suggestive of involvement in maintaining suture patency. Generation of BMP3 null mutants, however, has yielded little information regarding suture development. Aside from the observation of increased bone mineral density, no other significant abnormalities have been appreciated (55). Future work analyzing suture development in these BMP3 deficient mice may help elucidate whether BMP3-mediated antagonism is critical or redundant with existing Noggin pathways.

Apoptosis

The role of apoptosis in suture development remains largely unknown. Studies by Rice and colleagues have suggested a role for apoptosis in not only bone formation and resorption, but also in the normal processes of both suture fusion and patency (57). The precise function of apoptosis in suture fate, however, continues to remain controversial. Early work by Furtwangler et al. demonstrated apoptosis to occur among the leading osteogenic cells upon physical contact of adjacent heterotopic bone territories (58). This suggested an active role for programmed cell death in the maintenance of suture patency. Additional investigations on osteoblasts derived from patients with syndromic craniosynostosis demonstrated a significant reduction in the rates of apoptosis when compared to cells from normal controls; both annexin V and propidium iodide staining revealed diminished apoptotic rates in syndromic osteoblasts (59). Furthermore, coculture of normal cells with osteoblasts derived from synostosed sutures led to a reduction in the rate of programmed cell death for the normal osteoblasts (59). Finally, by manipulating levels of both (transforming growth factor) TGF- β 2 and TGF- β 3, Opperman and colleagues demonstrated increased numbers of apoptotic cells in sutures destined to remain patent (60). Integrating these studies, the data therefore suggest an active role for apoptosis in the maintenance of suture patency. By perturbing normal suture biology through reduction of programmed cell death, premature pathologic suture fusion may thus result.

Contrasting these findings, other investigations have demonstrated a relationship between craniosynostosis and upregulated apoptosis. Examining osteoblasts expressing gain-of-function FGFR-2 mutations, Mansukhani and colleagues found significantly elevated rates of programmed cell death relative to wild-type controls (61). Work by Lemonnier et al. likewise revealed increased apoptosis in osteoblasts harvested from patients with Apert syndrome when compared to normal osteoblasts (62). Western blot analysis in these Apert osteoblasts demonstrated increased levels of IL-1, Fas, and Bax protein, all known to be proapoptotic factors (62,63). Complementing these findings, studies with Nell-1 transgenic mice have similarly shown a relationship between suture fusion and apoptosis. Nell-1, a novel gene recently found to be upregulated in human unilateral coronal synostosis, has been demonstrated to be preferentially expressed in cranial intramembranous bone and involved in promoting osteoblast differentiation (64,65). Overexpression of Nell-1 in mice resulted in a significant increase in programmed cell death along the osteogenic fronts, as determined by terminal deoxynucleotidyl transferase mediated nick end labeling assays, and resultant coronal synostosis (65). The data thus suggest that increased apoptosis, whether through FGF signaling or enhanced Nell-1 expression, may play a role in premature suture closure. Despite these findings, however, the role of apoptosis in suture fate has yet to be fully defined. Future analysis

through inhibition of programmed cell death may help to further elucidate the interplay between apoptosis and premature, pathologic suture synostosis.

DISTRACTION OSTEOGENESIS

Repair and reconstruction of craniofacial skeletal hypoplasias including the mandible, maxilla, midface, orbits, and cranial vaults continues to represent a significant challenge to plastic surgeons. Traditional approaches for the repair of such dysmorphologies, including osteotomies and bone grafting, have been associated with both significant short- and long-term morbidities. Since the adoption of distraction osteogenesis to the craniofacial skeleton, however, this modality has rapidly become the treatment of choice for several bony deficiencies. Following the first report of human mandibular distraction by McCarthy in 1989, this technique has now become a standard tool for craniofacial surgeons to achieve dramatic midface and mandibular advancement (66).

Distraction osteogenesis, a powerful form of endogenous tissue engineering, achieves bone formation through the gradual separation of two osteogenic fronts. As described by Ilizarov, distraction typically employs rigid fixation with a several day latency period followed by gradual separation and stable fixation until radiographic and clinical assessment confirm the presence of mature, mineralized bone bridging the distraction gap (67–70). Despite increasing experience with this modality, however, surgeons nonetheless continue to confront a variety of troubling complications. Overall morbidity rates with distraction osteogenesis have been noted to be as high as 35%, with soft-tissue infection, osteomyelitis, pin-tract infection, scarring, and fibrous nonunion most commonly reported (71). Furthermore, patient discomfort and complications related to compliance undoubtedly add to reports of overall morbidity (71). Given these concerns, investigators have endeavored to define the mechanisms guiding successful bone formation in the distraction regenerate. These studies have specifically focused on both mechanobiology and molecular biology toward a goal of further optimizing clinical outcomes while minimizing associated complications.

Traditional approaches for the investigation of distraction osteogenesis have generally employed use of large animal models, including canine, ovine, and lupine species (72–75). Studies using such models have provided a foundation for the histologic and ultrastructural changes associated with robust bone formation (72–75). These early investigations, however, have been encumbered by animal size, cost, and availability of molecular reagents. Addressing these limitations, recent work by Fang and colleagues has established a mouse model for mandibular distraction osteogenesis, enabling access to the wide array of molecular reagents, microarray analysis, bioluminescent imaging, microcomputed tomography, and transgene constructs readily available in mice (76). The development of this model has allowed refinement in our understanding of distraction mechanics and biology, with clear advantages in cost, scalability, and flexibility, allowing for more detailed investigations.

Mechanobiology

Bone development and maintenance is highly dependent on mechanical environment, with dynamic loading critical for both preservation and increase of bone mass (77,78). Recent investigations have purported a role for tensile strain and hydrostatic stress in the coordination of multipotent mesenchymal tissue differentiation into bone, cartilage, fibrous tissue, and/or fibrocartilage (79–81). The definition of mechanical forces involved with bone deposition in the regenerate has therefore emerged as a specific area of interest for research on distraction osteogenesis. Work by Loba and colleagues have characterized the resultant stress and strain patterns during active mandibular distraction, showing typical strain ranging between 10% and 12.5% across the regenerate corresponding to periods of greatest bone formation (82). Upon further examination, strain was noted to have a viscoelastic response, with peak strain occurring immediately after distraction and then gradually diminishing to less than half of the maximum level with time (82). Specific patterns for these tensile strains and hydrostatic forces have been computed by finite element models. Moderate hydrostatic stress, consistent with intramembranous bone formation, was noted within the distraction gap whereas the

periosteal edges were calculated to experience mild compressive stress (82). This, in contrast, would be predictive of endochondral bone formation. Interestingly, these predictions based on finite element analysis have been shown to highly correlate with histologic data derived from numerous animal models of mandibular distraction (76).

With this blueprint in hand for the forces involved during distraction osteogenesis, recent studies have endeavored to manipulate the mechanical environment to hasten completion of successful bone formation. By attempting to minimize overall length of treatment, the necessity of a latency period has already come under question. Both animal and clinical reports have suggested no significant benefit for delay of distraction following osteotomy (71,83–85). While still practiced by most contemporary craniofacial surgeons, the use of a standard latency period may perhaps be expendable and its elimination may serve to shorten current protocols for distraction osteogenesis. Focus has also been directed toward mechanical callus stimulation to accelerate the period of consolidation. Studies have already implicated axial loading of long bone fracture segments in promoting healing and callus bulk (86). Adapting this principle to mandibular distraction, investigations have demonstrated cyclic loading of the regenerate during early consolidation to increase mineral apposition rate and radiographic cortical density (83). Alternative stimulation through pulse ultrasound has likewise been shown to accelerate development of a mineralized regenerate, with pro-osteogenic effects noted when daily low-intensity ultrasound was introduced (87,88). Whether through cyclic loading or pulsed ultrasound, callus stimulation thus offers another potential avenue to enhance bone formation in craniofacial distraction osteogenesis. Incorporating these findings with data derived from mechanical modeling, more efficient strategies for distraction osteogenesis may therefore be developed and translated to clinical practice.

Molecular Biology

With ongoing studies defining the mechanical forces involved in successful distraction, recent work has suggested an equal importance for a conducive biochemical environment to promote robust bone formation. With the development of a murine model for mandibular distraction, significant progress has been made in elucidating the molecular pathways involved for successful osteogenesis (76). Numerous studies in both rats and mice have suggested several factors, including BMPs and other members of the TGF- β superfamily, to be critically involved in bone formation within the distraction gap (89–91). Investigating BMP expression patterns in the regenerate, upregulation of BMPs 2, 4, and 7 were all noted during active mandibular distraction. In addition, chondrocytes were also found to increase BMP transcription, particularly during consolidation (91). Extending these findings, augmentation of BMP-2 levels during consolidation by means of adenoviral delivery led to dramatically increased bone deposition, thus suggesting one potential modality to promote bone formation and ultimate clinical outcomes (92).

Pro-angiogenic cytokines have similarly received significant attention with regard to successful bone formation during distraction osteogenesis. Fang et al. noted significantly increased levels of vascular endothelial growth factor (VEGF) and FGF-2 expression during periods of active distraction, with quantitative real-time RT-PCR demonstrating a fourfold increase in transcript levels (76). Analogous findings have also been reported in goat models of mandibular distraction, with intense immunohistochemical staining for both VEGF and FGF-2 noted during active distraction (93). Recent studies from our own laboratory have specifically focused on suppressing these angiogenic signals to demonstrate the importance of biomolecular environment in successful bone formation. Systemic administration of TNP-470, a fumagillin analogue capable of inhibiting endothelial cell proliferation and new capillary formation, resulted in complete fibrous nonunion following standard distraction (94). Histologic analysis of these fibrous regenerates revealed absence of new vessel formation, suggesting failure of angiogenesis may have contributed to the failure in bone formation noted (94). These data therefore underscore the interdependence between mechanical environment and angiogenesis for successful osteogenesis during distraction. Induction of pro-angiogenic cytokines, perhaps in concert with pro-osteogenic cytokines, may thus facilitate more rapid and robust bone deposition for improved clinical outcomes.

Integrating studies on distraction mechanics with investigations on cytokine biology, new insights into successful bone formation during distraction osteogenesis have thus emerged. With knowledge of forces engendered and biomolecular mechanisms involved, new protocols for craniofacial distraction may be developed and ultimately employed in the clinical setting to achieve robust bone deposition in the treatment of skeletal hypoplasias and other bony defects.

CELLULAR-BASED BONE TISSUE ENGINEERING

Though distraction osteogenesis represents a powerful tool for craniofacial repair, the mechanical principles underlying successful osteogenesis cannot be applied to every clinical situation in which bone generation is necessary. The push for alternative strategies has therefore led to the incorporation of autogenous, allogeneic, and prosthetic materials to reconstruct craniofacial defects (95–103). Despite the development of these methods to address repair of bony deficits, the multitude of approaches only serves to underscore the current limitations of clinical practice. Use of materials such as bone grafts, demineralized bone matrix, metal, glass, and polymethylmethacrylate are beset by numerous shortcomings including infection, immunologic rejection, graft versus host disease, and donor-site morbidity (102,104,105). Given these considerations, researchers have therefore sought to devise novel approaches to engineer bone in the craniofacial skeleton.

Combining recent advances in cellular and molecular biology with new insight into design and development of biocompatible scaffolds, the field of cellular-based tissue engineering has emerged as a potentially attractive approach for the generation of novel bone. At its core, multipotent or tissue-specific building blocks are combined with molecular and environmental cues to promote development of bone for craniofacial repair. Two broad fields of research have emerged from this paradigm investigating both the design of biodegradable scaffolds and the identification of an optimal cellular progenitor for bone engineering. Current work in these areas has made the use of cellular-based tissue engineering an attractive approach for repair of calvarial and facial osseous defects.

Scaffold Design

As the demand for modalities with which to engineer novel bone continues to endure, the need for a means of efficient osteoprogenitor delivery remains critical. Investigations have specifically endeavored to develop optimal biomimetic scaffolds to facilitate cellular delivery for three-dimensional tissue regeneration. Conceptually, these scaffolds should be biocompatible in addition to possessing the capacity for osteoinduction and controlled biodegradation while maintaining structural integrity. Ideally, such scaffolds would also allow for controlled delivery of cytokine signaling for orchestration of both proliferation and lineage-specific differentiation. Current scaffolds in use include natural, mineral-based, and synthetic polymers, each with their own advantages and disadvantages (102,103,106–110).

Natural scaffolds have routinely been employed in several clinical situations for both craniofacial and dental applications. Collagen and hyaluronic acid have been among the most frequently used scaffolds of this group (106,109). Studies in rats have demonstrated increased mandibular bone healing with the application of type I collagen scaffolds to the defects (106). Chitosan, another natural scaffold, has also been used in craniofacial repair, with studies showing improved healing in canine mandibular defects when this water-soluble form of chitin is injected into the regenerate (103). Because natural scaffolds lack structural rigidity, however, their use in load-bearing regions is severely limited. As an alternative to natural scaffolds, mineral-based scaffolds have been developed, primarily consisting of calcium phosphates in the form of hydroxyapatite and/or beta-tricalcium phosphate (111,112). By reproducing the chemical composition and structure of mature bone, mineral-based scaffolds impart an osteoinductive signal to coax progenitor cells down an osteogenic lineage. Both hydroxyapatite and beta-tricalcium scaffolds have been shown to promote ectopic bone formation when seeded with progenitor cells and implanted subcutaneously into immunodeficient mice (113,114). In addition, Schleiphake and colleagues have demonstrated calcium phosphate scaffolds to be capable of promoting repair in rat calvarial defects (115). Nonetheless, despite growing

acceptance of these materials in clinical use, their porous nature make these scaffolds quite brittle and prone to fracture in regions under mechanical load. Therefore, like natural scaffolds, mineral-based scaffolds lack the strength for use in regions without inherent stability.

Given these limitations, extensive research has focused on the development of synthetic scaffolds with greater durability. Current polymers available include polyglycolic acid, polylactic acid, polydioxanone, polycaprolactone, and various combinations of the above (110). The real advantage with synthetic scaffolds lies in their ability to be engineered with precise rates of resorption and mechanical strength. However, while their use in load-bearing regions is constructive, these scaffolds lack the osteoinductive properties enjoyed by both natural and mineral-based scaffolds. Recent work has therefore converged on the generation of hybrid scaffolds engineered with both mineral and synthetic components. Studies by Kokubo have capitalized on advances in mineralization techniques, with the development of novel scaffolds coated with biomimetic apatites for tissue engineering (116). These apatite-coated macroporous scaffolds combine the osteogenic properties of mineral-based scaffolds with the structural integrity and versatility of synthetic polymers. Investigations using such scaffolds in mice have already demonstrated their ability to promote healing of critical-sized calvarial defects when seeded with multipotent progenitor cells (117). With these advances in scaffold design, researchers have thus begun to define the optimal niche for the engineering of novel bone. Future work in the field of scaffold design will undoubtedly incorporate delivery of recombinant osteogenic factors to provide temporally and spatially modulated signals to coordinate more rapid cellular progression toward mature, differentiated bone.

Cell-Based Therapies

Investigations to define the consummate biologic building block for design of cellular-based therapies have been met with great interest given the promise for widespread application in craniofacial and orthopedic surgery. With their enormous biologic potential, human embryonic stem cells have received particular notoriety for potential use in tissue engineering (118). Research on these cells, however, remains fettered by multiple political and ethical hurdles making their use in therapeutic modalities illusory in the near term (119–121). Similarly, use of gene therapy and genetically modified adult cells continues to engender significant debate, given recent adverse clinical outcomes and calls for a potential moratorium (122–125). As an alternative resource, postnatal progenitor cells have recently emerged as an attractive candidate for use in cell-based tissue engineering strategies. With their ability to differentiate into multiple cell types and their relative abundance and availability, postnatal multipotent mesenchymal cells present an attractive candidate for use in craniofacial repair and reconstruction.

The majority of early work with postnatal progenitors has focused on mesenchymal stem cells naturally residing within bone marrow. Multiple studies have demonstrated the ability of these cells to develop along multiple lineage-specific paths including bone, cartilage, muscle, ligament, tendon, adipose, and stroma for regeneration of tissue throughout the body (126–128). *In vitro* studies by Pittenger and colleagues have revealed bone marrow-derived mesenchymal stem cells to possess the ability to specifically differentiate into fat, cartilage, and bone under appropriate culture conditions (126,129). Use of these cells to heal bony defects has been validated in several animal models, with healing of parietal defects in rabbits promoted by delivery of harvested mesenchymal stem cells within a fibrin glue construct to the region of injury (130,131). Similar investigations have demonstrated comparable applications for these multipotent cells in the reconstruction of porcine orbital defects (132). Despite these provocative findings, however, use of bone marrow-derived mesenchymal stem cells remains limited due to their selective serum requirements, their low frequency within the nucleated marrow cell fraction, and the donor-site morbidity associated with their harvest (133–136). Because of these concerns, investigators have sought different sources from which to obtain these postnatal multipotent progenitor cells for use in bone tissue engineering strategies.

Studies by Zuk and colleagues have been instrumental in the definition of a new adipose-derived mesenchymal cell (AMC) fraction as an alternative to their bone marrow counterparts (137,138). Unlike bone marrow-derived mesenchymal cells, AMCs are more readily accessible

and represent an available and easily expandable cellular source for tissue engineering applications. The acquisition of AMCs avoids many of the significant morbidities associated with bone marrow harvest and is not prone to whole blood contamination making its overall yield substantially greater (134). Investigations into characterizing the nature of these cells derived from human lipoaspirate have demonstrated a dynamic potential for the generation of fat, cartilage, muscle, and bone (137,138). Similar studies in mice have demonstrated an equally wide-ranging capacity for lineage-specific differentiation, with data also suggesting the possible retention of osteogenic potential irrespective of donor age (139).

The ability of adipose-derived multipotent mesenchymal cells to engineer bone *in vivo* has been well demonstrated by several recent investigations. AMCs harvested from Lewis rats have been shown to be able to form bone subcutaneously when implanted with polyglycolic acid grafts (140). Equivalent findings have also been noted with human AMCs, forming bone *in vivo* when seeded onto hydroxyapatite/tricalcium phosphate cubes and placed into severe combined immunodeficient mice (141). Furthermore, work by Dragoos and colleagues have recently demonstrated that processed lipoaspirate cells extracted from various human fat pads possess significant osteogenic potential when transfected with adenoviral delivered BMP-2 (142,143). Dramatic bone formation was noted when these transduced AMCs were implanted with collagen I matrices within a muscular compartment of mouse hind limbs (143). Focusing specifically on the craniofacial skeleton, work by our own laboratory has demonstrated mouse-derived AMCs to be capable of healing critical sized calvarial defects when implanted into the region of injury with apatite-coated poly (lactide-co-glycolide) scaffolds (117). Most intriguingly, however, preliminary use of AMCs has already been reported clinically, with repair of a large calvarial defect in a seven-year-old child (144). Using adipose-derived stem cells combined with harvested bone chips from the iliac crest, surgeons in Germany were able to demonstrate near complete continuity in the region of injury three months postoperatively (144). Considering these impressive results, significant progress has already been made with defining the ideal building block for cellular-based tissue engineering. Future work will continue to refine the use of AMCs and perhaps identify the optimal cellular subset within the heterogeneous mix of harvested cells for use in craniofacial bone tissue engineering.

SUMMARY

Craniofacial skeletal reconstruction remains a considerable biomedical burden considering burgeoning costs and the number of procedures performed annually to repair both congenital malformations and acquired bony injuries/defects. The need for effective strategies to treat these conditions has driven considerable research in the fields of craniosynostosis, distraction osteogenesis, and cellular-based bone engineering. With new insights into molecular biology, genetics, developmental biology, and biomedical/tissue engineering, investigators stand poised, over the next decade, to make further significant gains, indubitably culminating in the development of novel and more effective clinical approaches.

ACKNOWLEDGMENTS

This work was supported by the following grants: NIH R01 DE13194, R01 DE14526, the OAK Foundation to Michael T. Longaker, T90 DK070103 to Julia C. Chen, and the Ethicon-SUS Research Fellowship to Derrick C. Wan.

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about the book...

Addressing the complete range of craniofacial anomalies, from cleft lip and orthognatic surgery to acute facial fractures and tumors. **Craniofacial Surgery** provides step-by-step instruction on the anesthetic management, surgical work-up, and operative treatment of complex congenital or acquired anomalies affecting the head, upper face, and jaw. Written by seasoned experts who have developed a thorough clinical and basic knowledge in this evolving discipline, this source will comprehensively analyze basic areas of craniofacial surgery, and set the standard for the management of these challenging clinical entities.

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Printed in the United States of America

DK8210

ISBN 978-084938210-9



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